Cardiology in the Young

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Opening Hours

Day/Date		Secretariat	Exhibition
Wed.	16.5.07	07:30–18:30	15:00–20:00
Thu.	17.5.07	07:30–18:30	08:30–18:00
Fri.	18.5.07	07:30–18:30	08:30–18:00
Sat.	19.5.07	08:00–15:00	08:30–13:00

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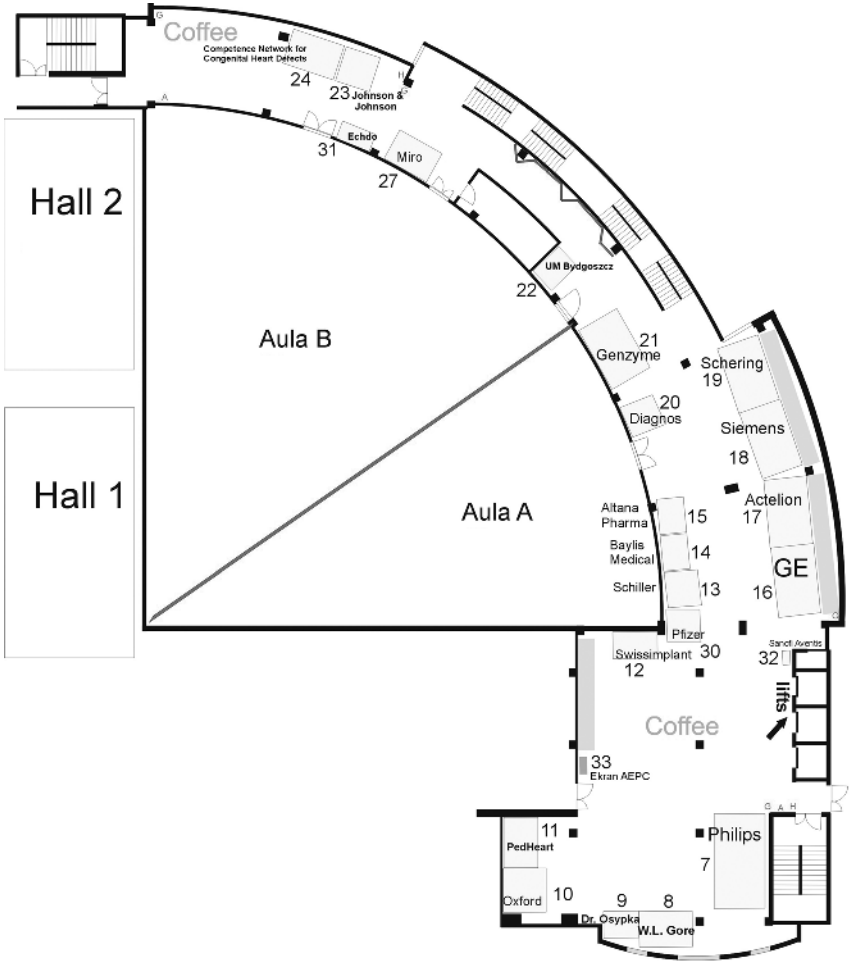
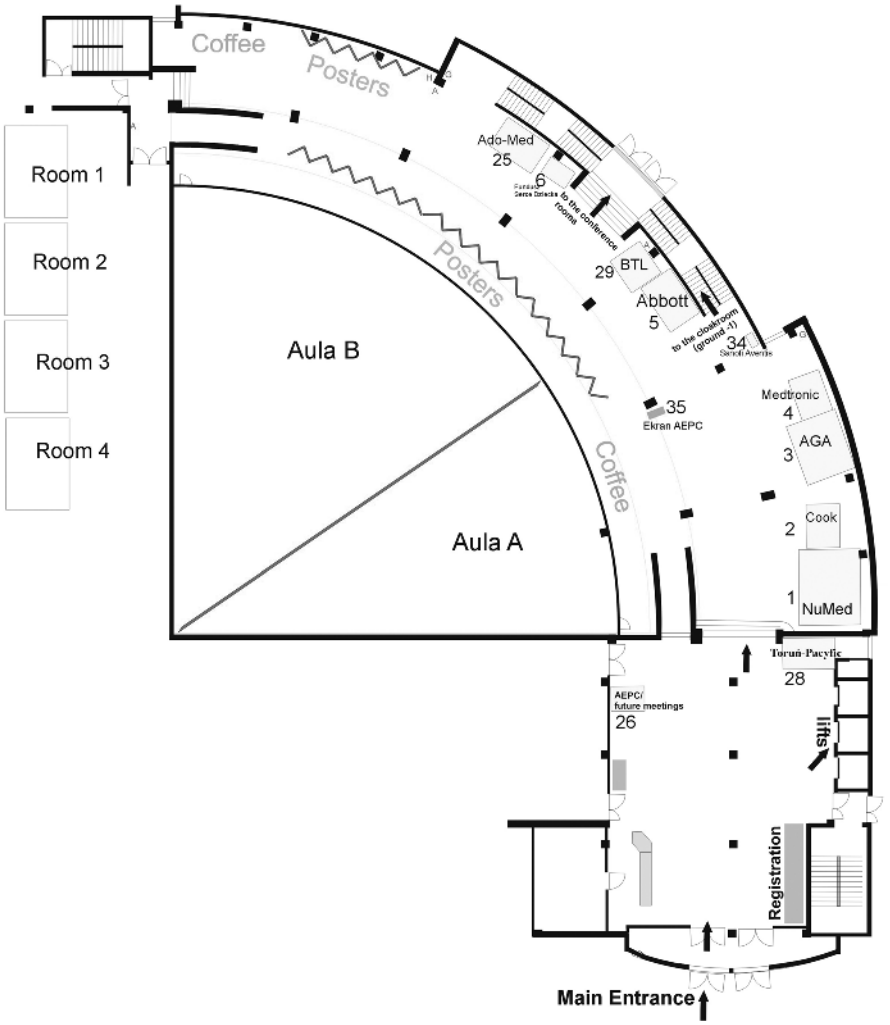


Conference Center of the Warsaw Medical Academy

Ground 1 Scale 1:250



Conference Center of the Warsaw Medical Academy

Ground 0 Scale 1:250

1. **CONFERENCE VENUE**
2. MARRIOTT
3. NOVOTEL CENTRUM
4. POLONIA PALACE
5. MERCURE F. CHOPIN
6. INTERCONTINENTAL
7. SOBIESKI
8. MDM
9. PREMIERE CLASE
10. NOVOTEL AIRPORT
11. **TYGMONT Club**
(Junior Member get together)

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Agenda AEPC Business Meeting Warsaw

Aula A, May 17, 2007

17.45–19.45

1. Welcome address of the President	Prof. Szatmári András
2. Obituary	
3. Report of the President	A. Szatmári
4. Report of the Secretary-General	E. Jokinen
5. Report of the Scientific Secretary	S. Qureshi
6. Report of the Treasurer	K. Schmidt
7. News from the Working Groups	
– Cardiac Dysrhythmias and Electrophysiolog	J. Janousek
– Fetal Cardiology	G. Tulzer
– Basic Genetics of Congenital Heart Defects	J. Parsons
– Pediatric Cardiovascular Intensive Care	E. Da Cruz
– Cardiac Imaging	E. Valsangiacomo Büchel
– Interventional Cardiology	M. Carminati
– Cardiovascular Morphology	A. Angelini
– Congenital Heart Surgery	G. Stellin
– Psycho-Social care from fetus to adult	U. Salzer-Muhar
– GUCH Task Force	F. Picchio
8. Report from the Advisory Board / National Delegates	S. Kaku
9. Introduction of New Members	E. Jokinen
10. Replacement of Council member	E. Jokinen
11. Election of the Scientific Secretary	E. Jokinen
12. Future Meetings	
2008 Padua / Venedig	G. Stellin
2009 during ESC–meeting	A. Szatmári
2010 Innsbruck	J. Stein
13. World Congress 2009	E. Jokinen
14. Handover of the presidency	A. Szatmári
15. Other businesses?	

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Wednesday, 16 May 2007				
	Aula A	Aula B	Hall 1	Hall 2
7:30				
8:00				
8:30				
9:00				
9:30				
10:00	08:30-15:00			
10:30				
11:00	Teaching Course on Grown Up			
11:30	Congenital Heart Disease			
12:00				
12:30				
13:00				
13:30				
14:00				
14:30				
15:00				
15:30				
16:00	16:00-17:00			
16:30	Opening Ceremony			
17:00	17:00-18:00			
17:30	Mannheimer Lecture			
18:00	18:00-20:00			
18.30	Welcome Reception			

Thursday, 17 May 2007				
	Aula A	Aula B	Hall 1	Hall 2
7:30				
8:00	08:00 - 09:00	08.00 - 09.00	08:00 - 09:00	08:00 - 09:00
8:30	WG Intervention - Business Meeting	WG Cardiac Imaging - Business Meeting	WG Intensive Care- Business Meeting	GUCH Task Force Business Meeting
9:00	09:00-10:30	09:00-10:30	09:00-10:30	
9:30	WG Intervention	WG Cardiac Imaging	WG Intensive Care	
10:00	Successes and failures in the Cathlab	Cardiac imaging in evolution – crystal ball gazing	Common issues in the daily management of acute paediatric cardiology patients	
10:30	10.40-11.45 moderated poster walk			
11:00				
11:30	11.45 - 12.45 State of the art. Hybrid approach to congenital heart defects			
12:00				
12:30	12.45 -13.45 lunch		12.45- 13.45	
13:00			Junior members "Future of Paediatric Cardiology: Generalist or Subspecialist?"	
13:30	13.45 - 14.45	13.45 - 14.45	13.45 - 14.45	13.45 - 16.45
14:00	Abstract session 1	Abstract session 2	Abstract session 3	
14:30	14.45 - 16.15	14.45 - 16.15		
15:00	Catheter closure of congenital VSD - fact or fantasy	**4D and Mutidimensional imaging - where do all the dimensions fit into for pediatrics ?**		Morphologic and CardioReplica demonstration
15:30	AGA	**General Electric**		
16:00	16.15 -16.45 Coffee break			
16:30	16.45 - 17.45	16.45 - 17.45	16.45 - 17.45	
17:00	Abstract session 4	Abstract session 5	Abstract session 6	
17:30	17.45 - 19.45 AEPC Business Meeting			
18:00				
18.30				
19.00				

Friday, 18 May 2007				
	Aula A	Aula B	Hall 1	Hall 2
7:30				
8:00	08:00 - 09:00	08:00 - 09:00	08:00 - 09:00	08:00 - 09:00
8:30	Fetal WG Business Meeting	Cardiac Dysrhythmias WG - Business Meeting	Congenital Heart Surgery WG - Business Meeting	Psycho-Social Care WG
9:00	09:00-10:30	09:00-10:30	09:00-10:30	
9:30	Fetal WG	Cardiac Dysrhythmias WG	Congenital Heart Surgery WG	
10:00	New Techniques in Fetal Cardiac Assessment	ICD therapy in children and patients with congenital heart disease	Management and Outcome of Patients with Pulmonary Atresia VSD and MAPCAs	
10:30	10.40 -11.45 moderated poster walk			
11:00				
11:30	11.45 - 12.45 State of the art. Fontan circulation. Late problems and future perspective			
12:00				
12:30	12.45 -13.45 lunch		**12.45 - 13.45 Genzyme symposium**	
13:00			**Pompe Disease and Treatment with Enzyme Replacement Therapy**	
13:30	13.45 -15.15	13.45 - 15.15		
14:00	Coarctation of aorta	**PAH: Translating evidence into clinical practice**		
14:30	Numed/Johnson&Johnson	**Actelion**		
15:00	15.15 - 16.15	15.15 - 16.15	15.15 - 16.15	
15:30	Abstract session 7	Abstract session 8	Abstract session 9	
16:00	16.15 - 16.30 Coffee break			
16:30	16.30 - 18.00	16.30 - 18.00		
17:00	**Latest advances in multidimentional and functional imaging in CHD**	**PAH in children: specific problems**		
17:30	Philips	Schering		
18:00				

Saturday, 19 May 2007				
	Aula A	Aula B	Hall 1	Hall 2
7:30				
8:00	08:00 - 09:00	08:00 - 09:00	08:00 - 09:00	
8:30	Basic Genetics WG - Business Meeting	Morphology WG - Business Meeting	AEPC Council Meeting - part I	
9:00	09:00-10:30	09:00-10:30		
9:30	Basic Genetics WG	Morphology WG		
10:00	Advances in the understanding of genetics and basic sciences in valvar congenital heart disease	Update on cardiomyopathies		
10:30	10.40 - 11.40	10.40 - 11.40	10.40 - 11.40	
11:00	Abstract session 10	Abstract session 11	Abstract session 12	
11:30	11.40 - 12.15 Coffee break			
12:00	12.15 - 13.15			
12:30	State of the Art: Arrhythmia			
13:00	13.30 - 14.30	13.30 - 14.30	13.30 - 14.30	
13:30	Abstract session 13	Abstract session 14	Abstract session 15	
14:00				
14:30				
15:00	15.00 post congress tour			

42nd Annual Meeting of
The Association for European Paediatric Cardiology
Warsaw Medical Academy Congress Center,
Warsaw/Poland, 16–19 May, 2007

FINAL SCIENTIFIC PROGRAMME

Time	Abstr. no.			Room
Tuesday, May 15, 2007				
09:00–15:00		**AEPC Council Meeting**	Marriot	Wars Room
15:00–16:30		**National Delegates Meeting**	Marriot	Sawa Room
16:30–18:00		**Professional Advisory Committee**	Marriot	Wars Room
16:30–18:00		**Scientific Advisory Committee**	Marriot	Sawa Room
		Teaching Course on Grown Up Congenital Heart Disease		
Wednesday, 16.05.2007				
07:30–08:30		Registration		
08:30–15:00		**Teaching Course on Grown Up Congenital Heart Disease**		Aula A
		Session 1: Transition from adolescent to adult life *Chairs: F. Picchio (Bologna IT), P. Hoffman (Warsaw, Poland)*		
08:30–08:35		**Opening remarks** *F. Picchio (Bologna IT)*		
08:35–08:50		**Transition from paediatric to adult life in patients with CHD** *P. Moons (Leuven, B)*		
08:50–08:55		**Discussion**		
08:55–09:10		**Pregnancy and contraception** *J. Stein (Innsbruck, AU)*		
09:10–09:15		**Discussion**		
09:15–09:30		**Coronary arteries in adults with CHD** *A. Bozio (Lyon, FR)*		
09:30–09:35		**Discussion**		
09.35–09:50		**Emergencies and sudden cardiac death in adults with CHD** *C. Wren (Newcastle, GB)*		
09:50–09:55		**Discussion**		
09:55–10:25		**Coffee Break**		
		Session 2: The aorta in adults with congenital heart disease *Chairs: E.J. Meijboom (Lausanne, CH), M. Yacoub (London, UK)*		
10:25–10:45		**Anatomico-pathological features of aortic involvement (Fallot, Marfan etc)** *A. Cook (London, UK)*		

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10:45–10:55	**Discussion**	
10:55–11:10	**Aortic valve disease** *F.J. Meijboom (Rotterdam, NL)*	
11:10–11:15	**Discussion**	
11:15–11:30	**Marfan syndrome** *M. Groenink (Amsterdam, NL)*	
11:30–11:35	**Discussion**	
11:35–11:50	**Surgical treatment** *B. Maruszewski (Warsaw, PL)*	
11:50–11:55	**Discussion**	
11:55–13:00	**Lunch**	
	Session 3: The failing heart in adults with congenital heart disease *Chairs: H. Kaemmerrer (Munich, DE), A. Szatmari (Budapest, HU)*	
13:00–13:15	**Heart failure in adults with CHD** *A. Bolger (London, UK)*	
13:15–13:20	**Discussion**	
13:20–13:35	**Treatment options for heart failure in adults with CHD** *F. Picchio (Bologna, IT)*	
13:35–13:40	**Discussion**	
13:40–13:55	**The failing right ventricle in adults with Tetralogy of Fallot** *F.J. Meijboom (Rotterdam, NL)*	
13:55–14:00	**Discussion**	
14:00–14:15	**The failing right ventricle in adults after atrial switch for TGA** *E.J. Meijboom (Lausanne, CH)*	
14:15–14:20	**Discussion**	
14:20–14:35	**Transplantation, who and when** *M. Yacoub (London, GB)*	
14:35–14:40	**Discussion**	
14:40–15:00	**Panel Discussion & Closing remarks** *A. Szatmari (Budapest, HU)*	

42nd Annual Meeting of AEPC

Wednesday, 16.05.2007

16:00–17:00	**Opening Ceremony** *Chairs: W. Kawalec (Warsaw, PL), A. Szatmari (Budapest, HU)*	Aula
17:00–18:00	**Mannheimer Lecture** *Chaisr: G. Brzezinska-Rajszys (Warsaw, PL), B. Maruszewski (Warsaw, PL)*	Aula
	Paediatric Cardiology in low income countries: our responsibilities *Dr Shakeel Qureshi (London, UK)*	
18:00–20:00	**Welcome Reception in the main foyer of the Congress Centre**	

Thursday, 17.05.2007

08:00–09:00	**AEPC Working Groups Business Meetings**	
	Interventional Cardiology Working Group	Aula A
	Cardiac Imaging Working Group	Aula B
	Pediatric Cardiovascular Intensive Care	Hall 1
	Business Meeting GUCH Task Force	Hall 2
09:00–10:30	**Working Groups Scientific Symposia**	
09:00–10:30	**Interventional Cardiology Working Group** **“Successes and failures in the catheter lab”** *Chairs: M. Carminati (Milan, IT), Dahnert Ingo (Leipzig, DE)*	Aula A

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Beware of the fragile aorta
R.P. Martin, M.S. Turner, S. Chakrabarti (Bristol, UK)
Late systolic murmur after perimembranous VSD closure
L. Mertens (Leuven, BE)
Complexities and complications in a Fontan circulation
F. Maymone-Martins, A. Texeira, R. Rossi, I. Menezes, R. Ferreira, R. Anjos, E. Dias da Silva, M. Abecais (Lisboa, PT)
Covered stent for late complications in a Fontan circulation
J. Kusa, M. Szkutnik, Bialkowski (Zabrze, PL)
Crossing the right hole
M. Witsenburg (Rotterdam, NL)
Hypotension after successful deployment of a Melody valve
D. Boshoff (Leuven, BE)

09:00–10:30 **Cardiac Imaging Working Group** Aula B
Cardiac imaging in evolution – crystal ball gazing
Chairs: W. Helbing (Rotterdam, NL), E. Valsangiacomo Buechel (Zurich, CH)
The future of echocardiography
L. Mertens (Leuven, BE)
Update in interventional magnetic resonance
R. Razavi (London, UK)
CT and congenital heart disease
H.J. Eichhorn (Heidelberg, DE)
Molecular imaging
O. Gamperli (Zurich, CH)

09:00–10:30 **Pediatric Cardiovascular Intensive Care Working Group** Hall 1
Common issues in the daily management of acute paediatric cardiology patients
Chairs: E. Lechner (Linz, AT), S. DeFilippo (Lyon, FR)

Hypoplastic Left Heart Syndrome: optimal pre-operative management
D. Schranz (Giessen, DE)
Emergency need for mechanical assistance. Methodology for an efficient and rapid ECLS setting
D. Metras (Marseille, FR)
Early vs. delayed extubation in the postoperative period of paediatric cardiac surgery: benefits, risk factors, safety criteria
E. da Cruz (Denver, USA)
Dexmedetomidine. Current experience in the management of the critical cardiac child
R. Munoz (Pittsburgh, USA)

10:40–11:45 **Moderated poster walk (session A)**
Chairs: A. Turska-Kmiec (Warsaw, PL), M. Mellander (Goteborg, SE)

10:40 **MPW-1** **Pathogenesis of Pulmonary Arteriovenous Malformations in children with congenital heart disease**
Watts G., Bharucha T., Clough G., Vettukattil J.J.
Congenital Cardiac Centre, Southampton, UK

10:46 **MPW-2** **Cryo-Ablation of Junctional Ectopic Tachycardia**
Snyder C. (1), Lucas V. (1), King T.(1), Darling R. (1), Young, M.L. (2), Bryant R.B. (3)
(1) Ochsner Clinic Foundation, Section of Pediatric Cardiology, New Orleans, LA, USA; (2) University of Miami / Jackson Memorial Medical Center, Section of Pediatric Cardiology, Miami, FL; (3) University of Florida, Division of Pediatrics, Jacksonville

10:52 **MPW-3** **Long-term follow up of bipolar steroid-eluting epicardial pacing leads in Children**
Tomaske M. (1), Gerittse B. (3), Kretzers L. (3), Pretre R. (2), Dodge Khatami A. (2), Rahn M. (2), Bauersfeld U. (1)

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Division of Paediatric Cardiology (1), University Children's Hospital Zurich, Switzerland; Division of Congenital Cardiovascular Surgery (2), University Children's Hospital Zurich, Switzerland; Medtronic Bakken Research Centre (3), Maastricht, The Netherlands

10:58 **MPW-4** **Short term follow up of implantable Cardioverter Defibrillator Systems using Epicardial and Pleural Electrodes in Pediatric Patients**

Bauersfeld U. (1), Tomaske M. (1), Dodge-Khatami A. (2), Rahn M. (2), Kellenberger C.J. (3), Pretre R. (2)
Division of Paediatric Cardiology (1); Division of Congenital Cardiovascular Surgery (2); Department of Diagnostic Imaging (3); University Children's Hospital, Zurich, Switzerland

11:04 **MPW-5** **Congenital heart block not associated with antiRo/SSA antibodies**

Vignati G. (1), Brucato A. (1), Milanesi O. (2), Grava C. (2), Pisoni M.P. (1), Martinelli S. (1), Canesi B. (1), Ruffatti (2)
(1) Niguarda Hospital, Milano, Italy, (2) Padova University, Padova, Italy

11:10 **MPW-6** **Assessment of cardiac function in severe twin to twin transfusion syndrome – multicenter study**

Dangel J. (3), Wloch A. (1), Respondek-Liberska M. (2), Czuba B. (1), Borowski D. (2), Cnota W. (1), Kaczmarek P. (2), Sodowski K. (1), Szaflik K. (2), Swiatkowska M. (4), Preis K. (4)
(1) Silesian Medical School, Katowice, Poland; (2) Polish Mother's Memorial Hospital, Lodz, Poland; (3) Medical University of Warsaw, Poland, (4) Medical University of Gdansk, Poland

11:16 **MPW-7** **Maternal Ingestion of Polyphenol-rich Common Beverages is Associated to Higher Fetal Ductus Arteriosus flow Velocities and Larger Right Ventricular size in Normal Pregnancies**

Zielinsky P. (1), Manica J.L., Piccoli Jr A.L., Nicoloso L.H., Frajndlich R., Menezes H.S., Busato A., Hagemann L., Moraes M.R., Silva J., Behrens T., Huber J., Silva M.B.
Fetal Cardiology Unit, Institute of Cardiology of Rio Grande do Sul/FUC, Porto Alegre, Brazil

11:22 **MPW-8** **Fetal Diagnosis of Cardiac Rhabdomyoma, its Management and Outcome**

Chockalingam P., Vial Y., Di Bernardo S., Pfammatter J.-P., Sekarski N., Mivelaz Y., Jeannet P.-Y., Meijboom E.J.
Centre Hospitalier Universitaire Vaudois, Lausanne (1), Div of Pediatric Cardiology, Inselspital, Bern (2), Switzerland

11:28 **MPW-9** **Missed Diagnoses in Antenatal Screening for Congenital Heart Disease: Implications for Training**

McBrien A., Casey F., Sands A., Craig B.
The Royal Belfast Hospital for Sick Children, Belfast, United Kingdom

11:34 **MPW-10** **Polish National Database for Fetal Cardiology at www.orpkp.pl**

Respondek-Liberska M. (1), Dangel J.H. (2), Wloch A. (3), Tobota Z. (4)
(1) Polish Mother's Memorial Hospital, Lodz, Poland; (2) Medical University of Warsaw, Poland, (3) Silesian Medical School, Poland; (4) Children's Memorial Health Institute, Warsaw, Poland

10:40–11:45 **Moderated poster walk (session B)**

Chairs: C. Wren (UK), M. Sadiq (Lahore, PK)

10:40 **MPW-11** **Novel Therapeutic Strategy for Eisenmenger Patients in Childhood**

Farahwaschy B., Gilbert N., Miera O., Hübler M., Ewert P., Berger F., Schulze-Neick I.
German Heart Institute Berlin, Germany

10:46 **MPW-12** **Pulmonary Blood Flow in Eisenmenger Patients Measured by Inert Gas Rebreathing**

Farahwaschy B., Gilbert N., Lunze K., Mebus S., Berger F., Schulze Neick I.
German Heart Institute Berlin, Germany

10:52 **MPW-13** **Cardiac function analysis with MRI: Assessment of measurement accuracy in a prospective multicenter study**

Kuehne T. (1)†, Gutberlet M. (2)†, Barth P. (3)§, Klimes K. (1), Froehlich C. (1), Franke D. (1), Kelter-Klöpping A. (3), Berger F. (1), Beerbaum P. (3)†,(†) princliple investigators, (§) postprocessing software development*

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Deutsches Herzzentrum Berlin, Department of Congenital Heart Diseases and Pediatric Cardiology (1); Humboldt University Berlin, Charité, Department of Radiology (2); Deutsches Herz- und Diabetiszentrum Bad Oeynhausen, Department of Pediatric Cardiology (3)

10:58 **MPW-14** **Effect of high-dose betablocker therapy on exercise capacity in patients with hypertrophic cardiomyopathy**
Bratt E.-L., de-Wahl Granelli A., Östman-Smith I.
The Department of Paediatric Cardiology, The Queen Silvia Children's Hospital, Göteborg, Sweden

11:04 **MPW-15** **Selection of high risk for death subgroups at presentation by classification tree analysis in infantile idiopathic dilated cardiomyopathy**
Azevedo V.M.P. (1), Santos M.A. (1), Castier M.B. (2), Amino J.G.C. (1), Cunha M.O.M. (1), Tura B.R. (1), Albanesi Filho F.M. (2), Xavier R.M.A. (1)
National Institute of Cardiology, Rio de Janeiro, Brazil (1); University of State of Rio de Janeiro, Rio de Janeiro, Brazil (2)

11:10 **MPW-16** **Long-term follow up of neoaortic regurgitation (AR) after the arterial switch operation for transposition of the great arteries**
Moll J. (1), Młudzik K. (1), Michalak K. (1), Kopala M. (2), Jarosik P. (2), Moll J. (2), Sysa A. (1)
Polish Mothers Memorial Hospital-Research Institute, Department of Cardiology Lodz, Poland (1); Polish Mothers Memorial Hospital-Research Institute, Department of Cardiosurgery, Lodz, Poland (2)

11:16 **MPW-17** **Role of Cardiac Troponin T in Clinically Suspected Acute Myocarditis in Children**
Sadiq M., Beg A., Rehman A.
The Children's Hospital and Punjab Institute of Cardiology, Lahore, Pakistan

11:22 **MPW-18** **Long-term outcome of pediatric patients with biopsy-proved myocarditis: comparison with late stage Kawasaki disease**
Yonesaka S. (1), Takahashi T. (2), Sato T. (2), Eto S. (2), Ueda T. (2), Sato A. (2), Otani K. (2), Ichinose K. (2), Kinjo M. (2)
(1) Department of Nursing, Hirosaki University School of Health Sciences, Hirosaki, Japan; (2) Department of Pediatrics, Hirosaki University School of Medicine, Hirosaki, Japan

11:28 **MPW-19** **Prevention of Respiratory Syncytial Virus Infection in Young Children With Heart Disease – A National Multicentric Study in a Mediterranean Country**
Moura C. (1), Carriço A. (1), Rebelo M. (5), Rossi R. (6), Marinho A. (4), Ramalheiro G. (3), Castela E. (3), Borges A. (8), Martins J. (7), Nunes M. (7), Loureiro M. (2), Alvares S. (2), Menezes I. (6), Pinto F. (5), Areias J. (1)
Hospital of S. João, Porto, Portugal (1), Hospital of Children Maria Pia, Porto, Portugal (2) Pediatric Hospital, Coimbra, Portugal (3) University Hospital, Coimbra, Portugal (4) Hospital of Santa Marta, Lisbon, Portugal (5) Hospital of Santa Cruz, Li

11:34 **MPW-20** **The Role of Telemedicine in the Remote Diagnosis of Congenital Heart Disease**
McCrossan B., Grant B., Morgan G., Sands A., Craig B. & Casey F.
Department of Paediatric Cardiology, Royal Belfast Hospital for Sick Children, UK

11:45–12:45 **State of the Art Session: Hybrid approach to congenital heart defects** Aula
Chairs: N.I Wilson (Oxford, UK), P. Burczynski (Warsaw, PL)

11:45–12:10 **Selection of types of procedures and patients for hybrid therapy**
J. Cheatham (Columbus, USA)

12:10–12:35 **Hybrid approach to HLHS**
D. Anderson (London, UK)

12:35–12:45 **Discussion**

12:45–13:45 **Lunch**

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12:45–13:45 **Junior members Lunch-break session "Future of Paediatric Cardiology: Generalist or Subspecialist?"** Hall 1

13:45–14:45 **Abstract Session 1: Imaging** Aula A

Chairs: E. Valsangiacomo Buechel (Zurich, CH), N. Blom (Leiden, NL)

13:45 **O1-1** **The Impact of Multiplanar Review of 3-Dimensional Echocardiographic Data on Management of Congenital Heart Disease**

Bharucha T., Roman K.S., Vettukattil J.J.
Congenital Cardiac Centre, Southampton, UK

13:55 **O1-2** **Evidence of liver fibrosis in children with Fontan circulation. Noninvasive assessement by a novel Doppler elastography and biochemical marker test**

Koch C. (1), Friedrich-Rust M. (2), Rentzsch A. (1), Fournier C. (2), Herrmann E. (2), Schwarz P. (1), Lindinger A. (1), Zeuzem S. (2), Schäfers H.J. (3), Abdul-Khaliq H. (1)
Department of Pediatric Cardiology, Saarland University Hospital, Germany (1); Department of Internal Medicine II, Saarland University Hospital, Germany (2); Department of Thoracic and Cardiovascular Surgery, Saarland University Hospital, Germany (3)

14:05 **O1-3** **Assessment of pulmonary artery size and function with phase contrast magnetic resonance imaging in patients after Fontan operation at young age**

Robbers-Visser D. (1,2), Helderman F. (3), Strengers J.L.M. (4), Kapusta L. (5), Dalinghaus M. (1), Bogers A.J.J.C. (6), Pattynama P.M.T. (2), Krams R. (3), Helbing W.A. (1,2)
Department of Pediatric Cardiology, Erasmus MC – Sophia Children's Hospital, Rotterdam the Netherlands (1); Department of Radiology, Erasmus MC, Rotterdam, the Netherlands (2); Department of Cardiology, Erasmus MC, Rotterdam, the Netherlands (3); Department

14:15 **O1-4** **Aortic root dysfunctioning and its effect on left ventricular function after the arterial switch operation assessed with MRI**

Grotenhuis H.B., Ottenkamp J., Fontein D., Hazekamp M.G., Vliegen H.W., Kroft L.C.M., de Roos A.
Leiden University Medical Center, Leiden, The Netherlands

14:25 **O1-5** **Noncompaction of the left ventricular myocardium in children: echocardiographic response to therapy**

Horowitz E.S., Huber J., Beherens T.
Instituto de Cardiologia do Rio Grande do SulPorto Alegre, Brazil

14:35 **O1-6** **MRI of aortic wall elasticity, aortic valve competence and LV function in patients with a non-stenotic bicuspid aortic valve**

Grotenhuis H.B., Ottenkamp J., Westenberg J.J.M., Bax J.J., Kroft L.J.M., de Roos A.
Leiden University Medical Center, Leiden, The Netherlands

13:45–14.45 **Abstract Session 2: General Paediatric Cardiology** Aula B

Chairs: E. Jokinen (Helsinki, FI), F. J. Meijboom (Rotterdam, NL)

14:55 **O2-1** **Caval flow reflects Fontan hemodynamics: Quantification by magnetic resonance imaging**

Ovroutski S., Klimes K., Ewert P., Alexi-Meskishvili V., Kühne T., Berger F.
German Heart Institute Berlin

15:05 **O2-2** **Arterialisation of hepatic blood flow in patients with a Fontan circulation**

Bryant T.J.C. (1), Burney K. (1), Stedman B. (1), Vettukattill J. (2), Haw M. (2), Salmon A. (2), Keeton B. (2), Cope R. (2), Hacking N. (1), Breen D.J. (1), Sheron N. (2), Veldtman G. (2)
Department of Radiology, Southampton General Hospital, Southampton, UK (1); Wessex Adult Congenital Heart Unit, Southampton General Hospital, Southampton, UK (2)

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15:15 **O2-3** **Compliance of the normal-sized aorta in adolescents with Marfan syndrome: Comparison of MR measurements of aortic distensibility and pulse wave velocity**
Eichhorn J.G. (1), Fink C. (2), Krissak R. (2), Rüdiger H.J. (1), Ley S. (2), Arnold R. (1), Kauczor H.-U. (2), Ulmer H.E. (1), Gorenflo M. (1)
University Children´s Hospital, Paediatric Cardiology, Heidelberg (1); DeutschesKrebsforschungszentrum (DKFZ), Radiology, Heidelberg (2); Ludwig-Maximilians-University, Radiology, Munich (3) Germany

15:25 **O2-4** **Strong and Independent Prognostic Value of Peak Circulatory Power in Adults With Congenital Heart Disease**
Giardini A. (1), Specchia S. (2), Berton E. (1), Sciarra F. (1), Coutsoumbas G. (2), Oppido G. (1), Gargiulo G. (1), Bonvicini M. (1), Picchio F.M. (1)
Pediatric Cardiology and Adult Congenital Unit, University of Bologna, Italy (1); Institute of Cardiology, University of Bologna, Italy (2)

15:35 **O2-5** **Predictors of rhythm and conduction disturbances in adult patients with congenital heart diseases**
Trojnarska O., Grajek S. (1) Kramer L. (2) Lanocha M. (1) Katarzynska A. (1)
University of Medical Sciences Department of Cardiology Poznan, Poland (1), University of Medical Sciences Department of Computer Science and Statistic (2)

15:45 **O2-6** **Health Related Quality of Life of Adults with Congenital Heart Defects**
Vigl M., Niggemeyer E., Busch U., Bauer U.
Kompetenznetz Angeborene Herzfehler, Berlin, Germany

13:45–14:45 **Abstract Session 3 General Paediatric Cardiology** Hall 1
Chairs: V. Pilossoff (Sofia, BL), F. Prada (Barcelona, SP)

13:45 **O3-1** **Effect of Beta-Blocker Therapy in Children with Dilated Cardiomyopathy: A Single Centre Retrospective Study**
Gagliardi M G., Pilati M., Saffirio C., Adorisio R., Sanders S.P.
Bambino Gesù Pediatric Hospital, Cardiology Department, Rome, Italy

13:55 **O3-2** **Utility of Signal-Averaged Electrocardiography in Pediatric ARVC**
Hamilton R. M., Kirsh J. A., Gross G. J., Basciano A., Stephenson E. A.
The Hospital for Sick Children, Toronto, Canada

14:05 **O3-3** **Quality of life after Fontan and TCPC-operation**
Nieminen H.P. (1), Mattila I.P. (1), Roine R.P. (2), Heikkilä A. (2), Sintonen H. (3), Sairanen H.I. (1)
Department of Surgery, Hospital for Children and Adolescents, Helsinki University Hospital, Helsinki, Finland(1), Group Administration, Helsinki and Uusimaa Hospital District, Helsinki, Finland (2), Department of Public Health and Finnish Office for Health (3)

14:15 **O3-4** **Low incidence of inappropriate or ineffective ICD therapy in children and adolescents with implantable cardioverter defibrillators. Results of a single center medium term study**
Botsch M., Will J.C., Opgen-Rhein B., Franzbach B., Berger F.
Department of Pediatric Cardiology, Charité Berlin, Germany

14:25 **O3-5** **Estimation of fetal atrioventricular (AV) time intervals by pulse Doppler and Doppler tissue echocardiography**
Dangel J.H., Hamela-Olkowska A., Wlasienko P.
Medical University of Warsaw, 2nd Department of Obstetrics and Gynecology, Perinatal Cardiology Department, Warsaw, Poland

14:35 **O3-6** **The Amplatzer® Membranous VSD Occluder and the Vulnerability of the Atrioventricular Conduction System**
Fischer G. (1), Apostolopoulou S.C.(2), Rammos S. (2), Schneider M.B. (3), Bjørnstad P.G. (4), Kramer H.H. (1)
Klinik für Kinderkardiologie, Universitätsklinikum Schleswig-Holstein, Campus Kiel, Germany (1); Department of Paediatric Cardiology, Onassis Cardiac Surgery Centre, Athens, Greece (2); Deutsches Kinderherzzentrum, Sankt Augustin, Germany (3) Paediatric Cardiology, Rikshospitalet – The National Hospital, University of Oslo, Oslo, Norway (4)

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13:45–16:45		**Morphologic and CardioReplica demonstration**	Hall 2
13:45–15:15		**Teaching session on Tetralogy of Fallot and Double Outlet Right Ventricle (CardioReplica)** *A. Juraszek (Boston, USA), S. Mottl-Link (Heidelberg, DE)* *T. Kuhne (Berlin, DE), G. Greil (London, UK)*	
15:15–16:45		**Cardiac Anatomy Demonstration** **Live video Specimens demonstration on Tetralogy of Fallot and Double Outlet Right Ventricle** *A. Angelini (Padua, IT)*	
14:45–16:15		**Educational unrestrictive grant sponsored by AGA Catheter closure of congenital VSDs – fact or fantasy** *Chairs: S. Qureshi (London,UK), B. Maruszewski (Warsaw, PL)*	Aula A
14:45–15:05		**What interventionalist should know before attempting VSD closure in catheter laboratory?** *A. Cook (London, GB)*	
15:05–15:25		**Are we really able to close surgically every VSD/VSDs with good short, mid and long-term result?** *B. Maruszewski (Warsaw, PL)*	
15:25–15:45		**What interventionalist can offer nowadays for VSD/VSDs closure?** *M. Carminati (Milano, IT)*	
15:45–16:05		**Should small perimembranous VSDs be closed?** *Tynan M. (London, UK)*	
16:05–16:15		**Summary and conclusion**	
14:45–16:15		**Educational unrestrictive grant sponsored by GE** **4D and Mutidimensional imaging – where do all the dimensions fit into for pediatrics?** *Chairs: L. Mertens (Leuven, BE), M. Vogt (Munich, DE)*	Aula B
		Quantitative Aspects of 2-D Strain and applications in Paediatrics: A Case-Based Approach *G. Di Salvo (Napoli, IT)* **Incorporating 4D Imaging Into Your Clinical Practice** *L. Mertens (Leuven, BE)* **CRT Dyssynchrony Assessment: how to evaluate? When to Use Velocity, Strain, and Speckle Tracking** *H. Abdul-Khaliq (Hamburg, DE)*	
16:15–16:45		**Coffee break**	
16:45–17:45		**Abstract Session 4: Fetal Cardiology** *Chairs: G. Sharland (London, UK), V. Fesslova (Milano, IT)*	Aula A
16:45	**O4-1**	**Autopsy protocol for prenatal cardiovascular anomalies – a substrate for successful cooperation between pathology, anatomy and prenatal cardiology specialists** *Kolesnik A. (1), Szymkiewicz-Dangel J. (2), Deregowski K. (3)* *Department of Anatomy, Medical University of Warsaw, Warsaw, Poland (1); Perinatology and Prenatal Cardiology Unit, Princess Anna Mazowiecka University Hospital, Warsaw, Poland (2); Pathology Laboratory, Princess Anna Mazowiecka University Hospital, Warsaw, Poland (3)*	
16:55	**O4-2**	**4D Fetal Echocardiography in Prenatal Diagnosis of Congenital Heart Disease** *Meijboom E.J., Chockalingam P., Mivelaz Y., Di Bernardo S., Sekarski N., Hohlfeld P. , Beurret Lepori N., Francini K., Vial Y.* *Centre Hospitalier Universitaire Vaudois, Lausanne, Switzerland*	
17:05	**O4-3**	**Ingestion of Herbal Teas and Grape Juice During Pregnancy is Associated to Fetal Ductal Constriction: a Clinical Approach** *Zielinsky P., Piccoli Jr A.L., Manica J.L., Nicoloso L.H., Menezes H.S., Frajndlich R., Petracco R., Busato A., Hagemann L., Moraes M.R., Silva J.*	

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Fetal Cardiology Unit, Institute of Cardiology of Rio Grande do Sul/FUC, Porto Alegre, Brazil

17:15 **O4-4** **Longitudinal Doppler study of the embryonic heart in low risk pregnacies and their follow up**
Wloch A. (1), Rozmus-Warcholinska W. (1), Czuba B. (1), Wloch S. (1), Cnota W. (1), K. Sodowski K. (1), Huhta J.C. (2)
Medical University of Silesia, Katowice, Poland (1) University of South Florida, St. Petersburg, United States (2)

17:25 **O4-5** **Comparison of first line transplacental antiarrhythmic therapies in fetal tachycardia**
De Groot E.E.C. (1), Blom N.A. (1), Clur S.A. (1), Rammeloo L. (1), Jaeggi E. (2)
Center of Congenital Heart Disease Amsterdam-Leiden (1), The Netherlands, Hospital for Sick Children, Toronto, Canada (2)

17:35 **O4-6** **3 Vessel View Z-score calculator: a practical tool for increasing detection of fetal coarctation of the aorta at the first screening examination**
Matsui H. (1), Mellander M. (1)(2), Pasquini L. (1), Seale A. (2), Roughton M. (3), Ho S.Y. (2), Gardiner H.M. (1)(2)
Faculty of Medicine, Imperial College at Queen Charlotte's & Chelsea Hospital, London, UK (1);Brompton Fetal Group, Royal Brompton Hospital, London, UK (2);Royal Brompton Hospital NHS Trust, London, UK (3)

16:45–17:45 **Abstract Session 5: End Stage Heart and Lung Disease** Aula B
Chairs: D. Schranz (Giessen, DE), W. Kawalec (Warsaw, PL)

16:45 **O5-1** **Outcomes in children with pulmonary arterial hypertension: Dutch Experience**
van Loon R.L.E. (1), Roofthooft M.T.R. (1), van Osch-Gevers M. (2), Delhaas T. (3), Strengers J.L.M. (4), Backx A. (5), Blom N.A. (6), Berger R.M.F. (1)
Department of Paediatrics, Divisions of Paediatric Cardiology, University Medical Center Groningen, University of Groningen, Groningen (1); Erasmus Medical Center Rotterdam, Rotterdam (2); University Hospital Maastricht, Maastricht (3); University Medical Ce

16:55 **O5-2** **Long-term effect of bosentan in patients with pulmonary arterial hypertension associated with systemic-to-pulmonary shunt: does the beneficial effect persist?**
van Loon R.L.E. (1), Hoendermis E.S. (2), Duffels M.G.J. (3), Vonk-Noordegraaf A. (4), Mulder B.J.M. (3), Hillege H.L. (2,5), Berger R.M.F. (1)
Department of Paediatrics, Division of Paediatric Cardiology (1), Department of Cardiology (2), and Department of Epidemiology-Research Coordination Center (5), University Medical Center Groningen, University of Groningen, Groningen; Department of Cardiology

17:05 **O5-3** **Decrease of interventricular Dyssynchrony after cardiac resynchronization therapy (CRT) in patients with congenital heart disease (CHD) is associated with improvement of ventricular function and physical performance**
Rita Schuck, Rentzsch A., Yegitbasi Y., Peters B., Miera O., Berger F., Abdul-Khaliq H.
Deutsches Herzzentrum Berlin. Berlin-Germany

17:15 **O5-4** **Outcome factors of idiopathic dilated cardiomyopathy in children. A follow-up review over twenty years**
Azevedo V.M.P. (1), Santos M.A. (1), Castier M.B. (2), Amino J.G.C. (1), Cunha M.O.M. (1), Tura B.R. (1), Albanesi Filho F.M. (2), Xavier R.M.A. (1)
National Institute of Cardiology, Rio de Janeiro, Brazil (1); University of State of Rio de Janeiro, Rio de Janeiro, Brazil (2)

17:25 **O5-5** **Time course of reverse remodeling after biventricular upgrade for conventional pacing induced left ventricular failure in congenital heart disease**
Janousek J. (1), Gebauer R.A. (2), Razek V. (1), Tomek V. (2), Kostelka M. (1)

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University of Leipzig, Heartcenter, Leipzig, Germany (1); Kardiocentrum, University Hospital Motol, Prague, Czech Republic (2)

17:35 **O5-6** **Biomarker responses during long-term mechanical circulatory support in children**
Heise G. (1), Lemmer J. (1), Weng Y. (2), Hübler M. (2), Alexi-Meskishvili V. (2), Böttcher W. (2), Hetzer R. (2), Berger F. (1), Stiller B. (1)
(1) Department of Congenital Heart Disease, (2) Department of Cardiothoracic and Vascular Surgery, Deutsches Herzzentrum Berlin, Germany

16:45–17:45 **Abstract Session 6: General Paediatric Cardiology** Hall 1
Chairs: S. Kaku (Lisbon, P), J. Stein (Innsbruck, AT)

16:45 **O6-1** **Multiplanar review of 3D Echocardiographic Datasets is an Accurate Method for Defect Sizing**
Bharucha T., Vettukattil J.J.
Congenital Cardiac Centre, Southampton, UK

16:55 **O6-2** **A Meta-Analysis of percutaneous versus surgical closure of ostium secundum atrial septal defects**
Butera G. (1), Biondi-Zoccai G. (2), Abella R. (1), Piazza L. (1), Chessa M. (1), Micheletti A. (1), Negura D. (1), Giamberti A. (1), Frigiola A. (1), Carminati M. (1)
Pediatric Cardiology – Policlinico San Donato IRCCS – Italy (1); Interventional Cardiology – University of Turin – Italy (2)

17:05 **O6-3** **Value of pharmacologic stress testing of so called "minimal" residual coarctation – it is time to change the traditional paradigma of good surgical results !**
Haas N.A., Schaeffler R., Beerbaum P., Laser T., Sarikouch S., Goerg R., Matthies W., Keceioglu D.
Heart and Diabetes Centre North-Rhine Westfalia, Bad Oeynhausen, Germany

17:15 **O6-4** **Growth among children with hypoplastic left heart syndrome (HLHS)**
Bijloo A.M., Rijlaarsdam M.E.B.
Leiden University Medical Center, Leiden, The Netherlands

17:25 **O6-5** **Impact of early and aggressive treatment with amiodarone on the therapeutic success and outcome in patients with postoperative tachyarrhythmias**
Haas N.A. (1,2), Camphausen C. (2), Kececioglu D. (1)
Heart and Diabtes Centre North-Rhine Westfalia, Bad Oeynhausen, Germany (1) The Prince Charles Hospital, Brisbane, Australia (2)

17:35 **O6-6** **Remote Magnetic Navigation: A new approach for mapping and ablation of supraventricular tachycardias in children and patients with congenital heart disease**
Pflaumer A. (1), Hessling G. (2), Luik A. (2), Ücer E. (2), Wu J. (2) Deisenhofer I. (2), Hess J. (1), Zrenner B. (2)
Pediatric Cardiology and Congenital Heart Disease (1), Cardiology (2), Deutsches Herzzentrum, Muenchen, Technische Universitaet Muenchen

17:45–19:45 **AEPC Business Meeting** Aula A

09:00–17:30 **Posters** Poster area

P-1 **Cryoablation versus Radiofrequency Ablation for AV Nodal Reentrant Tachycardia in Children and Adolescents**
Papagiannis J., Papadopoulou K., Kirvassilis G., Tsoutsinos A., Kantzis M., Laskari C., Kiaffas M., Apostolopoulou S., Rammos S.
Onassis Cardiac Surgery Center, Athens, Greece

P-2 **Long-term experiences of AutoCapture controlled pacing in children**
Tomaske M. (1), Harpes P. (3), Dodge-Khatami A. (2), Bauersfeld U. (1)

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Division of Paediatric Cardiology (1), University Children's Hospital Zurich, Switzerland; Congenital Cardiovascular Surgery (2), University Children's Hospital Zurich, Switzerland; Biostatistics Unit (3), University Zurich, Switzerland

P-3 **Heart rate variability and baroreflex sensitivity are related to fat amount in healthy children**

Czyz K. (1), Guzik P. (2), Bobkowski W. (1), Krauze T. (2), Piskorski J. (3), Surmacz R. (1), Siwińska A. (1)
Department of Pediatric Cardiology, Poznan University of Medical Sciences, Poznan, Poland (1); Department of Cardiology – Intensive Therapy, Poznan University of Medical Sciences, Poznan, Poland (2); Institute of Physics, University of Zielona Gora, Zie

P-4 **Morphologic and Functional Characteristics of the Cited2 Knockout Mouse Embryo: A Preliminary Study**

Huhta J.C., Serrano M.C., Acharya G., Han M., Linask K.K.
University of South Florida, Departments of Pediatrics and Obstetrics & Gynecology, USA

P-5 **A medium-chain triglyceride diet improves survival under stress but does not improve ventricular arrhythmias in mice with a fatty acid oxidation disorder**

Bartelds B. (1), Tokonuga N. (2), Yue Z. (2), Exil V. (2), Khuchua Z. (2), Kannankeril P.K. (2), Strauss A.W. (2)
Department of Pediatric Cardiology, Beatrix Childrens Hospital, University Medical Centre Groningen, The Netherlands (1); Division of Pediatric Cardiology, Department of Pediatrics, Vanderbilt University Medical Centre, Nashville, TN, USA (2)

P-6 **Deletion in the cardiac troponin I gene, resulting in premature stop codon, causes restrictive cardiomyopathy**

Sjöberg G., Kostareva A. (1), Gudkova A. (2), Morner S. (3), Semernin E. (2), Krutikov A. (2), Schlyakhto E. (2), and Sejersen T. (1)
Karolinska Institute, Stockholm, Sweden (1), Pavlov Medical University, St. Petersburg, Russia (2), University Hospital, Umea, Sweden (3)

P-7 **Computational/mathematical modeling of the systemic and pulmonary circulations with and without ventricular septal defect**

Corno A. (1), Milišić V. (2), Zunino P. (3), Quarteroni A. (3)
(1) Alder Hey Royal Children Hospital, Liverpool, England; (2) University Joseph Fourier, Grenoble, France (3) MOX, Milan, Italy

P-8 **Evaluation of 4 Methods of DNA Extraction from Formalin-Fixed, Paraffin-Embedded Human Heart Samples**

Lui H. (1, 2), Chalabreysse L. (1, 2, 3), Camminada C. (1, 2), Thivolet-Béjui F. (3), Bouvagnet P. (1, 2, 4)
(1) Laboratoire Cardiogénétique, Groupe Hospitalier Est, Hôpitaux de Lyon, Lyon, France, (2) Laboratoire Cardiogénétique, ERM 0107, INSERM, Lyon, France, (3) Laboratoire d'anatomo-pathologie, Groupe Hospitalier Est, Hôpitaux de Lyon, France, (4) Ser

P-9 **Correlation between PTPN11 gene mutation and congenital heart defect and the ECG pattern in Noonan syndrome**

Kapusta L., Croonen E.A., Burgt I. van de, Draaisma J.M.Th.
Radboud University Medical Centre, Nijmegen, The Netherlands

P-10 **Intravenous tezosentan and vardenafil equipotently reverse pulmonary vasoconstriction in experimental normobaric hypoxic pulmonary hypertension**

Geiger R. (1), Treml B. (2), Kleinsasser A. (2), Neu N. (3), Stein J.I. (1), Loeckinger A. (2)
Pediatric Cardiology (1), Anesthesiology and Critical Care Medicine (2), Pediatric Intensive Care (3); Medical University, Innsbruck, Austria

P-11 **Maternal Ingestion of Green Tea, Mate Tea and Grape Juice Cause Fetal Ductal Constriction: an Experimental Study**

Zielinsky P. (1), Areias J.C.N. (2), Piccoli Jr A.L. (1), Manica J.L. (1), Nicoloso L.H. (1), Menezes H.S. (1), Frajndlich R. (1), Busato A. (1),

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Petracco R. (1), Hagemann L. (1), Moraes M.R. (1), Silva J. (1), Allievi M. (1), Centeno P. (1), Abegg M. (1)
(1) Fetal Cardiology Unit, Institute of Cardiology of Rio Grande do Sul / FUC – ULBRA - UFRGS, Porto Alegre, Brazil; (2) Hospital São João, University of Porto, Portugal

P-12 **Left ventricular aneurysm in fetal life – a rare entity with an unpredictable outcome**

Fesslova V.M.E., Rosati E. (1), Boschetto C. (2)
Center of Fetal Cardiology, Policlinico San Donato, Milano, Italy; Dpt. of Ped. Cardiology, Ospedale A. Perrino, Brindisi, Italy (1); 1stt Obst. Gynec. Clinic, University of Milan, Italy (2)

P-13 **Diastolic function of the fetal heart assessing by tissue Doppler imaging: a comparison to premature infants and neonates**

Parezanovic V., Jovanovic I., Djukic M., Ilisic T., Ilic S., Vukomanovic G., Stefanovic I.
University Children's Hospital, Belgrade, Serbia

P-14 **Decreased serum insulin-like growth factor-I and insulin-like growth factor binding protein-3 associated with growth failure in patients with cyanotic heart disease**

Hallioglu O. (1), Alehan D. (2), Kandemir N. (3)
Mersin University Department of Pediatric Cardiology, Mersin, Turkey (1) Hacettepe University Department of Pediatric Cardiology (2) and Endocrinology (3), Ankara Turkey

P-15 **Clinical spectrum, morbidity, and mortality in 41 pediatric patients with ventricular noncompaction**

Olgunturk R. (1), Yildirim A. (1), Tunaoglu F.S. (1), Kula S. (1), Oguz D. (1), Sanli C. (1)
Gazi University Medical Faculty, Depertmant of Pediatric Cardiology, Ankara, Turkey

P-16 **Vascular endothelial growth factor, IL-6 and IL-8 levels in congenital heart disease**

Tunaoglu FS., Kula S., Zengin A., Olguntürk R., Saygılı A., Oğuz D.
Gazi University Medical School, Department of Pediatric Cardiology, Ankara, TURKEY

P-17 **A Large Pedigree With Mild And Severe Cases Of Congenital Heart Defects Segregates A Zic3 Missense Mutation**

Camminada C. (1, 2), Ollagnier C. (1, 2), Ducreux C. (3), Bozio A. (3), Bresson J.P. (4), Bouvagnet P. (1, 2, 3)
(1) Laboratoire Cardiogénétique, Groupe Hospitalier Est, Hôpitaux de Lyon, Lyon, France, (2) Laboratoire Cardiogénétique, ERM 0107, INSERM, Lyon, France, (3) Service de Cardiologie Pédiatrique, Groupe Hospitalier Est, Lyon, France, (4) Service de Gé

P-18 **Hospitalization for Respiratory Tract Infection in Young Children with Hemodynamically Significant Congenital Heart Disease: The 'CIVIC II' Study, Spain 2005-2006**

Medrano C., Garcia-Guereta L., Grueso J., Insa B., Ballesteros F., Casaldaliga J., Cuenca V., Escudero F., Garcia de la Calzada D., Luis M., Mendoza A., Carretero F., Rodríguez M.M., Suarez P., Quero C. On behalf of The CIVIC Study Group from the Spanish Society of Pediatric Cardiology and Congenital Heart Disease

P-19 **Can Preoperative Testing Predict Postoperative Outcome?**

Farahwaschy B., Lunze K., Mebus S., Berger F., Schulze-Neick I.
German Heart Institute Berlin, Germany

P-20 **Waist circumference and early detection of Metabolic syndrome**

Zavjalova L.G., Denisova D.V., Simonova G.I.
Institute of Internal Medicine, Novosibirsk, Russia

P-21 **Utility of a regional campaign for early diagnosis and treatment of Kawasaki's disease to prevent cardiac-coronary complications**

Bettuzzi MG., Colaneri M., Baldinelli A., Ricciotti R., Osimani P.
Presidio Cardiologico "G.M.Lancisi" – Ospedali Riuniti – Ancona - Italy

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P-22 **The Kurdistan Experience: Creating Paediatric Cardiac Services in the post – Saddam Era**
Arcidiacono C. (1), Tavormina R. (1), Gardi I. (2), Mohammed E.H. (3), Salih A.F. (4), Nuri H.A. (5), Shekho N.Q. (5), Aziz B.A. (6), Frigiola A. (1)
Policlinico San Donato IRCCS, Milan, Italy (1); Istituto Mediterraneo di Ematologia IRCCS, Rome, Italy (2); Azadi General Hospital, Kirkuk, Iraq (3); Paediatric Teaching Hospital, Sulaymaniyah (Kurdistan), Iraq (4), Hawler Medical University, Erbil (Kurdi)

P-23 **Comparison of clinical profile in children with different types of left ventricular hypertrophy in hypertrophic cardiomyopathy**
Ziolkowska L., Kawalec W., Pregowska K., Turska-Kmiec A., Tomyn-Drabik M., Mirecka-Rola A., Daszkowska J.
The Children's Memorial Heath Institute, Warsaw, Poland

P-24 **Metabolic cardiomyopathies in a paediatric population: clinical spectrum and outcome**
Alvares S., Bandeira A., Martins E., Andrade T., Loureiro M.
Hospital Crianças Maria Pia, Porto, Portugal

P-25 **Sexual life in male adolescents and young adults with congenital heart disease**
Antoniadis S., Dionyssopoulou E.
Diagnostic Center for Pediatric Cardiology Athens Greece

P-26 **Pulse-oximetry derived non-invasive Peripheral Perfusion Index as a possible tool for screening for congenital critical left heart obstruction: Comparison with reference values on 10 000 normal newborns**
de-Wahl Granelli A. (1), Östman-Smith I. (2)
Department of Clinical Sciences, Division of Paediatrics (1); The Sahlgrenska Academy, Göteborg University (2), Sweden.

P-27 **Frequency of diagnostic ECG-abnormalities in children of parents with familial hypertrophic cardiomyopathy**
Östman-Smith I., Bratt E-L., de-Wahl Granelli A.
Division of Paediatric Cardiology, Institute of Clinical Sciences, Queen Silvia Children's Hospital, Gothenburg, Sweden

P-28 **The QT and QTc intervals dispersion in sporting teenagers**
Dimitriu A.G. (1), Mihai F. (2), Pavel L. (1)
1. University of Medicine and Pharmacy Iasi Romania
2. University "Dunarea de Jos" Galati Romania

P-29 **Muscular Dystrophy and Cardiac Disease in Children**
Trigo C. (1), Paixão A. (1), Soudo A. (2), Kaku S. (1),
Hospital de Santa Marta Lisbon Portugal (1), Hospital de Dona Estefania Lisbon Portugal (2)

P-30 **Assessment of Relationship Between Selected Tissue Doppler Imaging Parameters of Left Ventricular Function and Systolic Blood Pressure Load in ABPM in Children After the Successful Repair of Aortic Coarctation**
Florianczyk T., Werner B.
Department of Pediatric Cardiology and General Pediatrics, The Medical University of Warsaw Warsaw, Poland

P-31 **Changes in left ventricular dimension during refeeding in anorexia nervosa associated with increased insulin-like growth factor 1 (IGF-1)**
Docx M.K.F. (1), Gewillig M. (2), Vandenberghe Ph. (1), Eyskens F. (3), Mertens L. (2)
Paediatric Cardiology Koningin Paola Kinderziekenhuis, Antwerp, Belgium (1); Paediatric Cardiology, University Hospital Gasthuisberg, Leuven,Belgium (2); Center of Metabolic Diseases Koningin Paola Kinderziekenhuis, Antwerp, Belgium (3)

P-32 **The role of exercise test in children after correction of congenital heart disease**
Wojcicka-Urbanska B., Werner B., Kucinska B., Florianczyk T.
Department of Pediatric Cardiology and General Pediatrics, The Medical University of Warsaw, Warsaw, Poland

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P-33 Changes in heart rate and blood pressure with refeeding in anorexia nervosa compared with endocrine parameters
Docx M.K.F. (1), Vandenberghe Ph. (1), Eyskens F. (2), Gewillig M. (3), Mertens L. (3)
Paediatric Cardiology Koningin Paola Kinderziekenhuis, Antwerp, Belgium (1); Center of Metabolic Diseases Koningin Paola Kinderziekenhuis, Antwerp, Belgium (2); Paediatric Cardiology University Hospital Gasthuisberg, Leuven, Belgium (3)

P-34 Long term postoperative results in isolated discrete subaortic stenosis
Kaneva-Nencheva A., Pilossoff V., Tzonzarova M.
National Heart Hospital, Sofia, Bulgaria

P-35 Sexual behaviour und erectile dysfunction in male patients with congenital heart disease
Hager A. (1), Vigl M. (2), Bauer U. (2), Köhn F.M. (3), Hess J. (1), Kaemmerer H. (1)
(1) Deutsches Herzzentrum München, TUM, Munich, Germany
(2) Kompetenznetz Angeborene Herzfehler, Berlin, Germany
(3) Andrologicum, Munich, Germany

P-36 Functional outcome in 9-13 year old children after surgical correction in the first year of life because of congenital heart defects
de Feijter A. (1), Beelen A. (1), Meester A. (1), Nollet F. (1), Ottenkamp J. (2)
Department of Rehabilitation, AMC, Amsterdam, the Netherlands (1)
CAHAL: Center for congenital Anomalies of the Heart Amsterdam/Leiden, the Netherlands (2)

P-37 Maximal aerobic capacity in children operated for congenital heart disease
Arvidsson D. (1), Sunnegårdh J. (2), Slinde F. (1), Hulthén L. (1)
Department of Clinical Nutrition, Sahlgrenska Academy at Göteborg University, Göteborg, Sweden (1); Queen Silvia Children´s Hospital, Sahlgrenska University Hospital, Göteborg, Sweden (2)

P-38 Outcome after aortic valve replacement with mechanical prosthesis in children and adolescents
Arnold R. (1), Ley S. (2),Loukanov T. (3), Sebening Chr. (3), Kleber B. (4), Ulmer H.E. (4), Hagl S. (3), Gorenflo M. (4)
Dept. Paed. Cardiology (1), University Medical Centre, Freiburg, i.Br.; Dept. Radiology (2), German Cancer Research Institute, Heidelberg; Dept. Cardiac Surgery (3) and Paed. Cardiology (4), University Medical Centre, Heidelberg, Germany

P-39 Centralized archiving of digital echocardiographic data for mid and long term multicenter studies within the competence net for congenital heart disease in Germany
Rentzsch A. (1), Böttler P. (2), Vogel M. (3), Sax U. (4), Müller S. (4), Lange P.E. (5), Bauer U. (5), Abdul-Khaliq H. (1)
Saarland University Hospital, Department for congenital heart disease/pediatric cardiology, Homburg (1); University Medical Center Freiburg, Department for pediatric cardiology and congenital heart disease (2); German Heart Center Munich (3); Georg-August

P-40 Adult patients after total correction of coarctation of aorta – evaluation of systolic and diastolic function of left ventricle by tissue dopler imaging
Trojnarska O., Lanocha M., Oko-Sarnowska Z., Szyszka A.
University of Medical Sciences in Poznan, Poland

P-41 Evaluation of Right Heart Myocardial Performance in Patients with Ebstein's Anomaly – a TDI Study
Hacke P. (1), Lunze F.I. (1), Nagdyman N. (1), Abd El-Rahman M.Y. (1) Abdul-Khaliq H. (3), Hetzer R. (2), Berger F. (1)
Deutsches Herzzentrum Berlin, Berlin, Germany: Abteilung für Angeborene Herzfehler/Kinderkardiologie (1), Abteilung für Herz-, Thorax- und Gefäßchirurgie (2) Universitätsklinikum des Saarlandes, Homburg/Saar, Germany: Klinik für Pädiatrische Kardiologie

P-42 Six-minute walk test with measuring wheel: reference values from 3 to 18 years of age

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Geiger R. (1), Strasak A. (2), Treml B. (3), Gasser K. (1), Kleinsasser A. (3), Stein J.I. (1), Loeckinger A. (3) Pediatric Cardiology (1), Medical Statistics, Informatics, Health Economics (2), Anesthesiology and Critical Care Medicine (3); Medical University, Innsbruck, Austria

P-43 **Clinical presentation and echocardiographic features of Double Orifice Mitral Valve**

Wójcik A., Klisiewicz A., Konka M., Szymanski P., Michalek P., Lusawa T., Rozanski J., Hoffman P.
Institute of Cardiology, Warsaw, Poland

P-44 **The geometry of the pulmonary artery bifurcation in children – an MRI in vivo study**

Valsangiacomo Buechel E.R., Knobel Z., Albisetti M., Bergsträsser E., Kellenberger C.J.
University Children's Hospital Zurich, Switzerland

P-45 **Tissue Doppler Imaging of infantile Pompe Patients shows regional systolic and diastolic Changes during long term Follow Up under Enzyme Replacement Therapy**

Schmidt D. (1), Hagel K.J. (1), Hahn A. (2), Schranz D. (1)
Pediatric Heart Center (1), Pediatric Neurology Center (2)
University Giessen, Germany

P-46 **Velocity vector imaging analysis of cardiac function in normal neonates**

Tsapakis E., De Catte L., Dermauw A., Boshoff D., Eyskens B., Gewillig M., Mertens L.
Department of Paediatric Cardiology, University Hospital Gasthuisberg, Leuven, Belgium

P-47 **Assessment of Left Heart Myocardial Performance in Patients with Ebstein's anomaly – a TDI study**

Hacke P. (1), Lunze F.I. (1), Nagdyman N. (1), Abd El-Rahman M.Y. (1), Abdul-Khaliq H. (3), Hetzer R. (2), Berger F. (1)
Deutsches Herzzentrum Berlin, Berlin, Germany: Abteilung für Angeborene Herzfehler/Kinderkardiologie (1), Abteilung für Herz-, Thorax- und Gefäßchirurgie (2), Universitätsklinikum des Saarlandes, Homburg/Saar, Germany: Klinik für Pädiatrische Kardiologi

P-48 **Interventricular septum function after early and late TOF correction, a Tissue Doppler Study**

Maschietto N. (1), Padalino M. (2), Pluchinotta F. (1), Stellin G. (2), Milanesi O. (1)
Paediatric Cardiology Unit, Department of Paediatrics, University Of Padua, Padova, Italy (1), Paediatric Cardiac Surgery Unit, Department of Cardiovascular surgery, University Of Padua, Padova, Italy (2)

P-49 **Impact, safety and hemodynamic effects of magnetic resonance imaging in pediatric cardiac intensive care patients**

Sarikouch S., Schaeffler R., Haas N.A., Kirchner G., Beerbaum P., Kececioglu D.
Department for Congenital Heart Disease, Heart and Diabetes-Center Northrine-Westfalia, Ruhr-University of Bochum, Bad Oeynhausen, Germany

P-50 **Comparison of Non-Invasive Cardiac Output Estimation Using Applanation Tonometry and Transmission Line Theory Versus Conventional Doppler**

Leung M.T. (1), Dumont G.A. (1), Potts J.E. (2), Sandor G.G.S. (2)
Department of Electrical and Computer Engineering (1), Children's Heart Centre, B.C. Children's Hospital (2), The University of British Columbia, Vancouver, Canada

P-51 **Perioperative Stent Implantation for Management of Pulmonary Artery Stenoses in Infants with Single Ventricle Physiology**

Sreeram N. (1), Emmel M. (1), Brockmeier K. (1), Hitchcock F. (2), Bennink G. (1)
University Hospital of Cologne, Cologne, Germany (1); University Medical Center, Utrecht, the Netherlands (2)

P-52 **Cor triatriatum dexter as a contraindication for transcatheter closure of secundum atrial septal defects with the Amplatzer Septal Occluder**

Konka M., Demkow M. (1), Ruyo W. (2), Hoffman P. (3)
Institute of Cardiology, Warsaw, Poland

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P-53 Outcome of Transcatheter Closure of Patent Ductus Arteriosus in Children Using the Amplatzer Duct Occluder

Thanopoulos B.D. (1), Rigby M.L. (2), Trapali Ch. (1), Karanasios E. (1), Eleftherakis N. (1), Hakim F.A. (3), Djukic M. (4), Simeunovic S. (4), Zarayelyan A. (5), Stefanadis Ch. (6)
'Aghia Sophia' Children's Hospital, Athens, Greece(1), Royal Brompton Hospital, London, UK, (2) 'Queen Alia'Heart Institute, Amman, Jordan, (3) University Children's Hospital, Servia-Montenegro (4), Medical State University Hospital, Yerevan, Armenia (5), H

P-54 Stent or Balloon for stenotic pulmonary arteries? Short and midterm results

Kantzis M. (1), Zartner P. (2), Wiebe W. (2), Apostolopoulou S.C. (1), Kaestner M. (2), Toussaint-Götz N. (2), Handke R.P. (2), Rammos S. (1), Schneider B.M. (2)
Department of Pediatric Cardiology, Onassis Cardiac Surgery Center, Athens, Greece (1); Department of Pediatric Cardiology, DKHZ Sankt Augustin, Germany (2)

P-55 Interventional catheterisation of systemic veins: early and long term results

Calcagni G. (1)(2), Marini D. (1), Gesualdo F. (2), Boudjemline Y. (1), Ou P. (1), Bonnet D. (1), Agnoletti G. (1)
(1) Necker Enfants Malades, Paris, France; (2) University La Sapienza, Rome, Italy

P-56 Hemodynamic assessment and types of interventions in symptomatic post-Fontan patients

Rudzinski A. (1), Werynski P. (1), Kordon Z. (1), Krol-Jawien W. (1), Zaluska-Pitak B. (1), Paruch K. (1), Kuzma J. (1), Kobylarz K. (2)
Department of Pediatric Cardiology, Collegium Medicum Jagiellonian University, Krakow, Poland (1), Department of Pediartic Anaesthesiology and Intensive Care, Collegium Medicum Jagiellonian University, Krakow, Poland (2)

P-57 National registry for treatment of coarctation of the aorta with stents

Brzezinska-Rajszys G. (1), Bialkowski J. (2), Sabiniewicz R. (3), Zubrzycka M. (1), Szkutnik M. (2), Ksiazyk J. (1), Kusa J. (2), Erecinski J. (3), Rewers B. (1), Kawalec W. (1)
Heart Catheterization Laboratory, The Children Memorial Health Institute, Warsaw, Poland (1), Congenital Heart Disease and Pediatric Cardiology Dpt, Silesian Center for Heart Diseases, Zabrze, Poland (2), Department of Paediatric Cardiology and Congenita

P-58 Catheterinterventions to reduce frequency of reoperation in patients after repair of common arterial trunc

Kaestner M. (1), Boscheinen M. (1), Toussaint-Götz N. (1), Grohmann J. (1), Wiebe W. (1), Kallenberg R. (1), Sinzobahamvya N. (2), Urban A.E. (2), Asfour B. (2), Schneider M.B.E. (1)
German Pediatric Heart Centre, Sankt Augustin, Germany, Dpt. Cardiology/Congenital Heart Desease (1); Dpt. Thoracic and Cardiac Surgery (2)

P-59 Percutaneous Treatment of Conduit and Pulmonary Arteries´ stenosis in modified Norwood procedure (Sano modification)

Zunzunegui J.L., Ballesteros F., Alvarez T., Camino M., Medrano M., Panadero E., Maroto E., Maroto C.
Gregorio Marañon Hospital, Madrid, Spain

P-60 Transcatheter closure of patent ductus arteriosus among native high-altitude habitants

Bialkowski J., Szkutnik M. (1), Menacho-Delgadillo R. (2), Palmero-Zilveti E. (2)
Silesian Center for Heart Diseases, Zabrze , Poland (1); Hospital Obrero, La Paz, Bolivia (2)

P-61 Taussig–Bing anomaly - late results after anatomic correction

Ostrowska K., Ostrowska K., Moll J.A., Binikowska J., Moll M., Dryek P., Sysa A., Moll J.J.
Polish Mother's Memorial Hospital, Lodz, Poland

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P-62 **Myocardial oxygenation and substrate metabolism in pediatric heart surgery: role of cardioplegia**
Åmark K. (1), Berggren H. (2), Björk K. (2), Ekroth A. (2), Ekroth R. (2), Sunnegårdh J. (1)
The Queen Silvia Children's Hospital and Department of Pediatrics (1), Department of Metabolic and Cardiovascular Research/Cardiothoracic Surgery (2), Göteborg University, Sweden

P-63 **Repair of congenital mitral valve anomalies in children: Long-term results in 52 cases**
Cikirikcioglu M. (1), Sierra J. (1), Kalangos A. (1)
University Hospital of Geneva, Department of Cardiovascular Surgery (1)

P-64 **The use of biodegradable rings in atrio-ventricular valve reconstruction**
Mrowczynski W. (1), Mrozinski B. (2), Wojtalik M. (1), Kalangos A. (3)
Dept. of Paediatric Cardiac Surgery – Poznan, Poland (1); Dept. of Paediatric Cardiology – Poznan, Poland (2); Clinique de Chirurgie Cardio Vasculaire – Geneva, Swtizerland (3)

P-65 **N-Terminal Pro-Brain Natriuretic Peptide as a Perioperative Biomarker in Paediatric Cardiac Surgery: Preliminary Results**
Freitas I., Nogueira G., Rebelo M., Afonso D.V., Kaku S., Fragata J.
Serviço de Cardiologia Pediátrica, Serviço de Cirurgia Cardiotorácica, Hospital de Santa Marta, Lisboa, Portugal

P-66 **Hemodynamic and oxygenation changes after oral Sildenafil for pulmonary hypertension in infants and children**
Doell C. (1), Dodge-Khatami A. (2), Fasnacht M. (3), Frey B. (1), Baenziger O. (1)
Divisions of Pediatric Intensive Care and Neonatology (1), Congenital Cardiovascular Surgery (2), Pediatric Cardiology (3), University Children's Hospital Zürich, Switzerland

Friday, 18.05.2007

08:00–09:00 **AEPC Working Groups Business Meetings**

Fetal Cardiology	Aula A
Cardiac Dysrhythmias and Electrophysiology	Aula B
Congenital Heart Surgery	Hall 1
Psycho-Social care from fetus to adult	Hall 2

09:00–10:30 **Fetal Cardiology Working Group** Aula A

New Techniques in Fetal Cardiac Assessment
Chairs: G. Tulzer (Linz, AT), J. Dangel (Warsaw, PL)

Imaging the fetal heart in 2007
J. Simpson (London, UK)

Functional assessment of the fetal heart using new techniques
C. Medrano (Madrid, SP)

Practical utility and limitations in diagnosing fetal heart defects
D. Bonnet (Paris, FR)

European study on fetal heart block: preliminary results
H. Eliasson (Stockholm, S)

09:00–10:30 **Cardiac Dysrhythmias and Electrophysiology Working Group** Aula B

ICD therapy in children and patients with congenital heart disease
Chairs: U. Bauersfeld (Zurich, CH), JM Happonen (Helsinki, FN)

Evolving ICD concepts and therapies in paediatric patients from shockbox to ICD-CRT
R. Hamilton (Toronto, CAN)

Indications for ICD therapy
T. Paul (Göttingern, DE)

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ICD implant techniques
C. Berul (Boston, USA)

ICD follow-up and complications
U. Bauersfeld (Zurich, CH)

09:00–10:30 **Congenital Heart Surgery Working Group** Hall 1

Management and Outcome of Patients with Pulmonary Atresia VSD and MAPCAs
Chairs: H. Sairanen – Helsinki (Finland)
O. Stumper – Birmingham (UK)

Morphology and embryology of complex pulmonary atresia
A. Cook (London, UK)

Investigation and intervention in complex pulmonary atresia
O. Stumper (Birmingham, UK)

Surgical strategies for complex pulmonary atresia
J.V. Comas (Madrid (SP)

The Stanford experience and outcomes with complex pulmonary atresia
V.M. Reddy (Stanford, USA)

10:40–11:45 **Moderated poster walk (session A)**
Chairs: J. Ksiazyk (Warsaw, PL), F. Picchio (Bologna, IT)

10:40 **MPW-21** **Evaluation of the influence of left lateral thoracotomy on scoliosis development in children with aortic coarctation**
Sabiniewicz R., Roclawski M., Potaz P., Erecinski J.
Department of Paediatric Cardiology and Congenital Heart Disease Medical University Gdansk, Poland

10:46 **MPW-22** **Assessment of left ventricular size and function using 3D-echo generated volume-time-curves in small infants with severe left ventricular outflow tract obstruction**
Herberg U. (1), Krötz A. (1), Breuer T. (2), Schmitz C. (3), Breuer J. (1)
Pediatric Cardiology, University of Bonn (1); Technische Informatik, FH Bonn Rhein Sieg, St. Augustin (2); Pediatric Cardiac Surgery, University of Bonn (3), Germany

10:52 **MPW-23** **Long term results of early total surgical repair of tetralogy of Fallot**
Dasheva A., Pilossoff V., Lazarov S., Mitev P., Christov G., Tzonsarova M.
National Heart Hospital, Sofia, Bulgaria

10:58 **MPW-24** **Plasma levels of B-type natriuretic peptide at mid- and long-term follow-up after total cavopulmonary connection**
Koch A., Zink S., Singer H.
Paediatric Cardiology, Hospital for Children and Adolescents, University of Erlangen-Nürnberg, Germany

11:04 **MPW-25** **Ablation Success and Long Term Outcome in Catheter Ablation of Atrial Tachycardias in Patients after the Fontan Operation**
Pflaumer A. (1), Haimerl M. (1), Hessling G. (2), Deisenhofer I. (2), Estner H. (2), Zrenner B. (2), Hess J. (1)
Pediatric Cardiology and Congenital Heart Disease (1), Cardiology (2), Deutsches Herzzentrum Muenchen, Technische Universitaet Muenchen

11:10 **MPW-26** **Transcatheter closure of congenital ventricular septal defects in a GUCH: mid term results and complications**
Chessa M., Butera G., Carminati M., Negura D., Piazza L.., Bossone E., Bussadori C., Micheletti A., Arcidiacono C.
Pediatric Cardiology and Adult with Congenital Heart Defect, IRCCS-Policlinico San Donato Milan-Italy

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11:16 **MPW-27** **Effects of Percutaneous Closure of Secundum Atrial Septal Defect in Adults on Ventricular Morphology and Function Examined by Tissue Doppler Echocardiography**
Jakubowska E. (1), Demkow M. (2), Kowalik E. (1), Lusawa T. (1), Konka M. (1), Hoffman P. (1)
1. Congenital Heart Disease Department, National Institute of Cardiology Warsaw, Poland. 2. Ist Hemodynamic Department, National Institute of Cardiology Warsaw, Poland

11:22 **MPW-28** **Predictors of ventricular arrhythmia in adult patients with congenital heart diseases**
Trojnarska O., Grajek S. (1), Kramer L. (2), Katarzynska A. (1), Lanocha M. (1)
1. University of medical Sciences Department of Cardiology
2. University of Medical Sciences Department of computer Science and Statistic Poznan, Poland

11:28 **MPW-29** **Social Aspects of the Life of Adults with Congenital Heart Defects**
Vigl M., Niggemeyer E., Busch U., Bauer U.
Kompetenznetz Angeborene Herzefehler, Berlin, Germany

11:34 **MPW-30** **Quality of life and exercise capacity in 497 patients with congenital heart disease**
Hager A., Gratz A., Hess J.
Department of Pediatric Cardiology and Congenital Heart Disease, Deutsches Herzzentrum München, TUM, Munich, Germany

10:40–11:45 **Moderated poster walk (session B)**
Chairs: D. Hagler (Rochester, US), K. Schmidt (Dusseldorf, DE)

10:40 **MPW-31** **Ventricular repolarisation versus transvalvular gradient and left ventricular mass in children with aortic valve stenosis**
Piorecka-Makula A., Werner B., Florianczyk T.
Department of Pediatric Cardiology and General Pediatrics, The Medical University of Warsaw, Warsaw, Poland

10:46 **MPW-32** **Transcather closure of congenital and acquired ventricular septal defects with Amplatzer devices**
Bialkowski J., Szkutnik M., Kusa J.
Congenital Heart Disease and Pediatric Cardiology Dept., Silesian Center for Heart Diseases, Zabrze, Poland

10:52 **MPW-33** **Repeat percutaneous pulmonary valve implantation: proof of principle as long-term management strategy for RVOT conduit dysfunction**
Nordmeyer J. (1), Lurz P. (1), Coats L. (1), Khambadkone S. (1), Frigiola A. (1), Cullen S. (1,2), Yates R. (1), Bonhoeffer P. (1,2)
UCL Institute of Child Health and Great Ormond Street Hospital for Children, London, UK (1) The Heart Hospital, London, UK (2)

10:58 **MPW-34** **Transesophageal echocardiography in newborns with miniaturized transducer to guide interventional cardiac catheterization**
Ballesteros F., Zunzunegui J.L., Alvarez T., Maroto E., Riaño B., Camino M., Panadero E., Medrano C., Maroto C.
Gregorio Marañon Hospital, Madrid, Spain

11:04 **MPW-35** **Evaluation of residual shunt at mid-long term follow-up of percutaneous closure of patent foramen ovale**
Lunardini A., Ait-Ali L., Amoretti F., Pratali L.**, Kristo I., Quatrinil., Spadoni I.*
*Pediatric Cardiology and GUCH Unit, *Radiology, CNR, "G. Pasquinucci Hospital", Massa, **CNR, Institute of Clinical Physiology, Pisa, Italy*

11:10 **MPW-36** **Interventional closure of fenestration as a final stage of treatment in HLHS patients after Fontan operation**
Mazurek-Kula A. (1), Moszura T. (1), Dryzek P. (1), Ostrowska K. (1), Moll J.A. (1), Moll J.J. (2), Sysa A. (1)
Cardiology Department Polish Mother's Memorial Hospital, Research Institute (1) Cardiosurgery Department Polish Mother's Memorial Hospital, Research Institute (2) Lodz, Poland

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11:16 **MPW-37** **Transposition of the great arteries and left ventricular outflow tract obstructions – is there a place for switch operation?**
Santos M.A., Azevedo V.M.P., Amino J.G.C., Cunha M.O.M., Tura B.R., Xavier R.M.A.
National Institute of Cardiology – Rio de Janeiro – Brazil

11:22 **MPW-38** **Long term evaluation of coronary arteries in patients after the anatomical correction of transposition of the great arteries**
Moll M., Moszura T., Mludzik K., Moll J.A., Dryzek P., Sysa A., Moll J.J.
Polish Mother's Memorial Hospital, Lodz, Poland

11:28 **MPW-39** **Pre- and post cardiac surgery weight gain of infants under 1 year of age in relation to neurodevelopmental outcome**
Knirsch W. (1), Zingg W. (2,3), Bernet V. (4), Beck I. (5), Bauersfeld U. (1), Latal B. (5)
Divisions of Pediatric Cardiology (1), Pediatrics (2), Neonatology and Pediatric Intensive Care (4), Child Development Center (5), University Children´s Hospital Zurich, Switzerland Hospital Epidemiology (3), University Hospital of Geneve, Switzerland

11:34 **MPW-40** **Dexmedetomidine, a Novel Agent for the Acute Treatment of Supraventricular Tachyarrhythmias after Pediatric Cardiac Surgery**
Chrysostomou C., Shiderly D., Berry D., Morell V., Munoz R.
Children's Hospital of Pittsburgh – University of Pittsburgh, USA

11:45–12:45 **State of the Art Session:** Aula
Fontan circulation – Late problems and future perspectives
Chairs A. Szatmari (Budapest, HU), D. Schranz (Giessen, DE)

11:45–12:10 **Lessons learnt and solutions**
G. Stellin (Padua, IT)

12:10–12:35 **Late complications in Fontan patients**
M. Gewillig (Leuven, BE)

12:35–12:45 **Discussion**

12:45–13:45 **Lunch**

12:45–13:45 **Lunch break symposium Pompe Disease and Treatment with Enzyme Replacement Therapy (satellite symposium sponsored by Genzyme)** Hall 1
Chairs: A. Tylki-Szymanska (Warsaw, PL)

Pompe Disease: a continuum of clinical phenotypes
A. Tylki-Szymanska (Warsaw, PL)

Advancements in the diagnosis of Pompe Disease
Z. Lucaks (Hamburg, DE)

Cardiac manifestations of Pompe Disease and treatment results with ERT (Myozyme™)
E. Cerini (Mantova, IT)

13:45–15:15 **Educational unrestrictive grant sponsored by Numed/Johnson & Johnson** Aula A
Coarctation of the aorta
Chairs: F. Berger (Berlin, DE), M. Chessa (Milano, IT)

13:45–14:05 **Coarctation of the aorta – hypoplasia of the aortic segments. What are we dealing with?**
A. Cook (London, UK)

14:05–14:25 **Coarctation of the aorta – surgery. Is it really a piece of cake?**
W. Brawn (Birmingham, UK)

14:25–14:45 **What interventionalists can offer instead of or after surgery**
G. Brzezinska-Rajszys (Warsaw, PL)

14:45–15:05 **Coarctation of the aorta – one stage intervention or whole life treatment?**
J. Vriend (Amsterdam, NL)

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15:05–15:15 **Summary and conclusion**

13:45–15:15 **Satellite symposium sponsored by Actelion** Aula B
Pulmonary arterial hypertension: Translating evidence into clinical practice
Chairs: M. Beghetti (Geneva, CH), R. Berger (Groningen, NL)

Challenging convention: Management of PAH-CHD including Eisenmenger Syndrome
M. Beghetti (Geneva, CH)

Under close examination: The right ventricle in PAH
R. Berger (Groningen, NL)

Applying the evidence in everyday clinical practice
D. Bonnet (Paris FR), M. Beghetti (Geneva, CH), R. Berger (Groningen, NL)

Discussion

15:15–16:15 **Abstract Session 7: Arrhythmias** Aula A
Chairs: T. Paul (Gettingen, DE), A. Turska-Kmiec (Warsaw, PL)

15:15 **O7-1** **The utility of ECG criteria in Pediatric ARVC**
Hamilton R. M., Iori S., Buffo I.
The Hospital for Sick Children, Toronto, Canada

15:25 **O7-2** **Predictors of Response to Cardiac Resynchronization Therapy (CRT) in Pediatric and Congenital Heart Disease. Subanalysis of a Retrospective European Multicenter Study**
Janousek J. (1), Grollmuss O. (1), Abdul-Khaliq H. (2), Gebauer R.A. (3), Rosenthal E. (4), Villain E. (5), Fruh A. (6), Blom N.A. (7), Happonen J.-H. (8), Bauersfeld U. (9), Jacobsen J.R. (10), Bink-Boelkens M.T. (11), Delhaas T. (12), Papagiannis J. (13), Trigo C. (14)
Dept. of Pediatric Cardiology, University of Leipzig, Heartcenter, Leipzig, Germany (1); Clinic for Congenital Heart Defects and Pediatric Cardiology, Deutsches Herzzentrum Berlin, Berlin, Germany (2); Kardiocentrum, University Hospital Motol, Prague, Czech Rep.

15:35 **O7-3** **Lack to Prove Hemodynamic Short-Term Benefit of Biventricular Pacing in Patients with d-TGA after Atrial Switch**
Schweigmann U., Schermer E., Engl G., Geiger R. Stein J.I.
University Hospital Innsbruck, Department of Pediatrics III Cardiology, Allergology and Cystic Fibrosis, Austria

15:45 **O7-4** **Implantable cardioverter defibrillator therapy in children: long term clinical outcome and risk factors for shocks**
Heersche J. (1), Bink-Boelkens M.T. (2), Ten Harkel A.D.J. (3), Clur S.A. (1), Strengers J. (4), Reimer A. (5), Blom N.A. (1)
Center for Congenital Heart Disease Amsterdam-Leiden (1), University Medical Center Groningen (2), Erasmus Medical Center (3), University Medical Center Utrecht (4), University Medical Center Nijmegen (5), The Netherlands

15:55 **O7-5** **Cryoablation at growing myocardium: Results of intracoronary ultrasound and coronary angiography after energy application**
Kriebel T. (1), Hermann H.-P. (2), Schneider H. (1), Kroll M. (1), Sigler M. (1), Paul T. (1)
Department of Pediatric Cardiology and Pediatric Intensive Care Medicine, Georg-August-University Göttingen, Germany (1) Department of Cardiology and Pneumology Georg-August-University Göttingen, Germany (2)

16:05 **O7-6** **Catecholaminergic Polymorphic Ventricular Tachycardia in Pediatrics**
Snyder C. (1), Moltedo J. (2), Salerno J. (3), Bryant R. (4), Cannon B. (5)
Ochsner Clinic Foundation, New Orleans LA, USA (1); FLENI Institute, Buenos Aires, Argentina (2), Seattle Children's Hopsital, Seattle WA, USA (3), University of Florida, Jacksonville FL, USA (4), Texas Children's Hosptial, Houston, TX, USA (5)

15:15–16:15 **Abstract Session 8: Interventional Cardiology** Aula B
Chairs: P. Ewert (Berlin, DE), J. Cheatham (Columbus, USA)

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15:15 **O8-1** **Immediate haemodynamic effect and short term clinical follow up following percutaneous pulmonary valve implantation**
Lurz P. (1), Nordmeyer J. (1), Coats L. (1), Schievano S. (1), Frigiola A. (1), Khambadkone S. (1), Cullen S. (2), Walker F. (2), Taylor A. M. (1), Bonhoeffer P. (1,2)
1. UCL Institute of Child Health and Great Ormond Street Hospital, London for Children, UK, 2. Department of Adult Congenital Heart Disease, The Heart Hospital, London, UK

15:25 **O8-2** **Medium term results of stent implantation for pulmonary artery branch stenosis in infants and small children**
Van Esch E., Bökenkamp R., Hazekamp M.G., Clur S.A., Rammeloo L., Ottenkamp J., Blom N.A.
Center of Congenital Heart Disease Amsterdam-Leiden, The Netherlands

15:35 **O8-3** **ASD and PFO closure with the Solysafe device First clinical experience**
Ewert P. (1), Dähnert I. (2), Hess O. (3), Schuler G. (2), Sick P. (4), Sievert H. (5), Söderberg B. (6)
German Heart Institute, Berlin, Germany (1); Heart Institute, Leipzig, Germany (2); Universitätsspital Bern, Bern, Switzerland (3); Krankenhaus der Barmherzigen Brüder, Regensburg, Germany (4); Cardiovascular Center, Frankfurt, Germany (5); Queen Silvia C

15:45 **O8-4** **Biocomaptibility of atrial septal defect closure devices: Immunohistochemical characterisation of neo-tissues**
Foth R., Jux C., Kriebel T., Sigler M.
Department of Paediatric Cardiology and Intensive Care, Göttingen University, Germany

15:55 **O8-5** **Stent implantation in the arterial duct from the axillary artery in duct dependent pulmonary blood flow: anatomical and technical aspects**
Michel-Behnke I., Akintuerk H., Hagel K.J., Valeske K., Schranz D.
Pediatric Heart Centre Giessen, Germany

16:05 **O8-6** **Outcome of Patent Ductus Arteriosus Stenting in Patients with Ductal Dependent Pulmonary Circulation**
Hussain A., Al-Zharani S., Arfi M.A., Al-Ata J., Galal M.O.
King Faisal Specialist hospital & Research Centre, Jeddah, Saudi Arabia

15:15–16:15 **Abstract Session 9: Surgery** Hall 1
Chairs: J. Moll (Lodz, PL), S. Kaku (Lisbon, PT)

15:15 **O9-1** **Open heart surgery in neonates and premature infants weighing less than 2.5 kg**
Lechner E., Hofer A., Mair R., Sames-Dolzer E., Steiner J.J., Vondrys D., Tulzer G.
Children´s Heart Center Linz, Austria

15:25 **O9-2** **The Effect of Ductal Diameter on Surgical and Medical Closure of Patent Ductus Arteriosus in Preterm Neonates: Size Matters**
Dodge-Khatami A. (1), Tschuppert S. (1), Doell C. (2), Arlettaz R. (3), Baenziger O. (2), Rousson V. (4), Prêtre R. (1)
Congenital Cardiovascular Surgery (1), Pediatric Intensive Care (2), Neonatology (3), Department of Biostatistics (4), University Children's Hospital, Zurich, Switzerland

15:35 **O9-3** **Performing Ross earlier – a chance to avoid autograft dilatation and valve failure in the follow-up?**
Kopala M., Moll J.A., Mudzik K., Moll J.J.
Polish Mother's Memorial Hospital, Lodz, Poland

15:45 **O9-4** **Perventricular, Non Surgical Aortic Reconstruction**
Sideris E., Bramos D., Sideris B., Calahanis M., Christianakis E., Moulopoulos S.
Athenian Institute of Pediatric Cardiology, Athens, Greece

15:55 **O9-5** **Twenty year review: Arterial Switch Operation cardiac angiography follow-up at OLHSC**
Prendiville T., Duff D., Oslizlok P., Walsh K.
Our Lady's Hospital for Sick Children, Ireland

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16:05 **O9-6** **ECMO-Transport: For the safe transport of critical ill pediatric patients**
Reckers J., Asfour B., Hraska V., Haun C., Fink C.
German Pediatric Heart Center Sankt Augustin, Sankt Augustin, Germany

16:15–16:30 **Coffee break**

16:30–18:00 **Industry satellite symposium sponsored by Philips** Aula A

Latest Advances in Multidimensional and Functional Imaging in Congenital Heart Disease: Echocardiography, MRI and Cardiovascular X-ray
Chairs: J. Simpson (London, UK), G. Brzezinska-Rajszys (Warsaw, PL)

LIVE 3D Echo – do we really need it?
J. Simpson (London, UK)

CMRI – clinical application in congenital heart disease – now and tomorrow
T.Kühne (Berlin, DE)

Rotational Angiography – its' possible role in congenital heart disease
P. Ewert (Berlin, DE)

16:30–18:00 **Educational unrestrictive grant sponsored by Schering** Aula B

Pulmonary arterial hypertension in children: specific problems
Chairs: M. Beghetti (Geneva, CH), I. Schulze-Neick (Berlin, DE)

Etiology and epidemiology
M. Beghetti (Geneva, CH)
PAH prognostic markers in children
R. Berger (Groningen, NL)
Can we use ESC guidelines in children? Diagnostic paediatric algorithm
I. Schulze-Neick (Berlin, DE)
Can we use ESC guidelines in children? Therapeutic paediatric algorithm
D. Schranz (Giessen, DE)

09:00–17:30 **Posters** **Poster area**

P-67 **Do threshold trend fluctuations of epicardial leads predict pacing and sensing characteristics?**
Tomaske M. (1), Harpes P. (3), Dodge-Khatami A. (2), Amacker N. (1), Bauersfeld U. (1)
Division of Paediatric Cardiology (1), University Children's Hospital Zurich, Switzerland; Division of Congenital Cardiovascular Surgery (2), University Children's Hospital Zurich, Switzerland; Biostatistics Unit (3), University Zurich, Switzerland

P-68 **Impantable Cardioverter Defibrillators Safety and Efficacy in Children and Young Adults with Congenital Heart Disease**
Papagiannis J., Kantzis M., Kirvassilis G., Evgeniadou E., Sarris G.E., Rammos S.
Onassis Cardiac Surgery Center, Athens, Greece

P-69 **Anti-inflammatory Effects of HMG-CoA Reductase Inhibitors (Statins) on Acute Coronary Arteritis in a Rabbit Model of Kawasaki disease**
Ozawa S., Hamaoka K.
Kyoto Prefectural University of Medicine Graduate School of Medical Science, Kyoto, Japan

P-70 **Role of single domains of the Staphylococcal adhesin fibronectin-binding protein A: involvement in invasion and activation of human vascular endothelial cells in a model of S. aureus experimental endocarditis**
Heying R. (1,3), van de Gevel J. (3), Que Y.A. (2), Moreillon P. (2), Beekhuizen H. (3)
Department of Pediatric Cardiology, Childrens University Hospital, Düsseldorf, Germany (1); Laboratory of Infectious Diseases, University Hospital of Lausanne, Lausanne, Switzerland (2); Department of Infectious Diseases, LUMC, Leiden, The Netherlands (3)

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P-71 **Persistent of high body mass index in pre-pubertal obese children is an early determinant of Pulse Wave Velocity elevation, a marker of arterial stiffness**
Aggoun Y., Farpour-Lambert N.J., Beghetti M.
Paediatric Cardiology Unit, Geneva, Switzerland

P-72 **Expression of two-pore domain channel genes in TASK-1 deficient and wild type mice quantified by real time polymerase chain reaction**
Donner B.C., Hüning A., Schmidt K`.G.
Department of Pediatric Cardiology and Pneumology, Heinrich Heine University Duesseldorf, Germany

P-73 **Moderate hypothermia suppresses inflammatory response and cellular activities in human endothelial cells**
Schmitt K.R.L. (1), Diestel A. (2), Abdul-Khaliq H. (3), Berger F. (1,2)
Deutsches Herzzentrum Berlin, Germany (1), Charité Universitätsmedizin Berlin, Germany (2), Klinik für Pädiatrische Kardiologie Universitätsklinikum des Saarlandes, Homburg, Germany

P-74 **Cytokines, matrix metalloproteinases and tissue inhibitors of metaloproteinases in children after cardiac surgery**
Pasnik J. (1), Cywiska-Bernas A. (1), Mazurowski W. (1), Zeman K. (1), Moll J.A. (2), Sysa A.(2), Moll J.(3)
Department of Pediatrics, Prevention of Cardiology and Clinical Immunology, Medical University Lodz, Poland (1), Department of Pediatric Cardiology of Institute of Polish Mother's Health Lodz, Poland (2), Department of Pediatric Cardiosurgery, Institute o

P-75 **Stent implantation in Aortic Coarctation patients is not related with an increased risk of exercise induced hypertension**
Teixeira A., Ferreira Santos J., Mendes M., Ferreira R., Maymone Martins F., Anjos R.
Hospital de Santa Cruz, Pediatric Cardiology, Carnaxide, Portugal

P-76 **Inhaled Iloprost Therapy in Children with Pulmonary Arterial Hypertension**
Beghetti M. (1), Doran A.K. (2), Mallory G. (3), Barst R.J. (4) Law Y. (5), Abman S.H. (2), Ivy D.D. (2)
Children's University Hospital, Geneva, Switzerland (1), The Children's Hospital, Denver, USA, (2), Texas Children's Hospital, Houston Texas, USA (3), Columbia University College of Physicians and Surgeons and New York Presbyterian Hospital, New York, USA

P-77 **Prenatal echocardiographic diagnosis of congenital heart disease: quid prodest?**
Grison A. (1), Cerutti A. (1), Biffanti R. (1), Pluchinotta F.R. (1), Bissoli G. (1), Padalino M. (2), Stellin G. (2), Milanesi O. (1)
Department of Pediatric (1); Department of Pediatric Cardiac Surgery, University of Padova-School of Medicine. Padova, Italy

P-78 **Tissue Doppler Imaging indices of systolic and diastolic function in the normal fetal heart**
Dangel J.H., Hamela-Olkowska A., Wlasienko P.
Medical University of Warsaw, 2nd Department of Obstetrics and Gynecology, Perinatal Cardiology Department, Warsaw, Poland

P-79 **Bruce Treadmill Test in Children: First Normal Values for Children from Western Europe**
Mivelaz Y., Di Bernardo S., Meijboom E.J., Fall A.L., Sekarski N.
Pediatric Cardiology, Cardiovascular and Metabolic diseases Center, Lausanne Children's University Hospital (CHUV), Switzerland

P-80 **Serum cardiac Troponin T levels in active carditis due to acute rheumatic fever**
Oguz D. (1), Atmaca E. (2), Ocal B. (2), Karakurt C. (3), Sungur M. (4), Karademir S. (5)
Gazi University Ankara-Turkey (1); Sami Ulus Children' Hospital Ankara-Turkey (2); Inonu University Malatya-Turkey (3); 19 Mayis University Samsun-Turkey (4); Suleyman Demirel University Isparta-Turkey (5)

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P-81 **The influence of cardiosurgery procedures on the lymphocyte subpopulations in children with congenital heart defects operated in first year of life**
Cywiska-Bernas A. (1), Paśnik J. (1), Banasik M. (2), Moll J.A. (3), Sysa A. (3), Moll J. (4), Zeman K. (1)
Department of Pediatrics, Preventive Cardiology and Clinical Immunology, Medical University, Lodz, Poland (1); Department of Clinical Immunology, Institute of Polish Mother's Health, Lodz, Poland (2); Department of Pediatric Cardiology, Institute of Polis

P-82 **Assessment of left ventricular systolic function with Tissue Doppler Imaging and conventional echocardiographic methods in children after the successful repair of aortic coarctation**
Florianczyk T., Werner B.
Department of Pediatric Cardiology and General Pediatrics, The Medical University of Warsaw, Warsaw, Poland

P-83 **Acute heart disease following Chikungunya virus infection in the neonate**
Attali T. (1), Ramful D. (1), Chuong V. (1), Bonnet D. (2)
Neonatal intensive care unit-CHD Felix Guyon-Saint-Denis de la Réunion - France (1); Pediatric Cardiology, Necker-Enfants Malades, Paris V, AP-HP, Paris-France (2)

P-84 **Register of congenital heart defects (CHD) in Croatia (Croatian study - October 2002–January 2007)**
Malcic I., Dilber D., Mustapic Z, Kniewald H., Saric D.
Department of paediatric cardiology, Clinical Hospital Centre Zagreb, Croatia P-85 - withdrawn

P-86 **A rare cause of mitral involvement in the children: the Geleophysic Dysplasia (GD)**
Marini D. (1), Cormier-Daire V. (2), Agnoletti G. (1), Vouhé P. (3), Bonnet D. (1)
Pediatric Cardiology, Université Paris V, Necker, Paris (1), Medical Genetic, Université Paris V, Necker, Paris (2), Pediatric Cardiac Surgery, Université Paris V, Necker, Paris (3)

P-87 **Children with Congenital Heart Disease: Effectiveness of a Respiratory Synctial Virus Immuno-Prophylaxis Program**
Human D.G., Crosby M.C., Cender L.M., Potts J.E., Sandor G.G.S.
Children's Heart Centre, B.C. Children's Hospital and The University of British Columbia, Vancouver, Canada

P-88 **N-terminal-pro-B natriuretic peptide levels in patients after Fontan operation correlate with congestive heart failure**
Lechner E., Gitter R., Mair R., Schreier-Lechner E., Vondrys D., Tulzer G.
Children´s Heart Center Linz

P-89 **Sildenafil in Cystic Fibrosis as a Disease Model for Pulmonary Arterial Hypertension Caused by Impaired Gas Exchange**
Mebus S. (1), Kleemann R. (2), Miera O. (1), Berger F. (1), Schulze-Neick I. (1)
German Heart Institute Berlin, Germany (1); Carl-Thiem-Klinikum Cottbus, Germany (2)

P-90 **Early detection of myocardial dysfunction in childhood patients with beta-thalassaemia major**
Ucar T. (1), Ileri T. (2), Uysal Z. (2), Atalay S. (1), Tutar E. (1)
Ankara University School of Medicine, Department of Pediatric Cardiology (1), Department of Pediatric Haematology (2), Ankara, Turkey

P-91 **Preoperative mortality in patients with transposition of the great arteries**
Ten Harkel A.D.J., Dalinghaus M., Helbing W.A.
Erasmus MC-Sophia, Rotterdam, The Netherlands

P-92 **Diagnostic value of noninvasive scintigraphy with 99mTc-Anti-Granulocyte antibody in children with clinically suspected myocarditis – a preliminary report**
Ziolkowska L., Kawalec W., Biernatowicz M., Kaminska A., Turska-Kmiec A., Boruc A., Czarnowska E., Pronicki M., Tomyn-Drabik M., Brzezinska-Rajszys G.
The Children's Memorial Health Institute, Warsaw, Poland

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P-93 High Rate of Off-Label use in cardiovascular paediatric pharmacotherapy requires new focus in research
Laer S. (1), Hsien L. (1), Breddemann A. (1), Frobel A.K. (1), Heusch A. (2), Schmidt K.G. (2)
Clinical Pharmacy and Pharmacotherapy, Heinrich-Heine-University of Düsseldorf, Germany (1), Paediatric Cardiology and Pneumology, Heinrich-Heine-University of Düsseldorf, Germany (2)

P-94 Peculiar cardiac involvement in the infant of diabetic mothers
Dimitriu A.G. (1), Iliescu R. (2), Stamatin M. (2), Pavel L. (1)
1. Ist Clinic of Pediatrics, University of Medicine and Pharmacy Iasi Romania
2. Department of Neonatology, University of Medicine and Pharmacy Iasi Romania

P-95 Congenital complete AV block in children - clinical course and outcome
Pavlova M., Kaneva A., Marinov R., Tzonzarova M.
National Heart Hosptal, Sofia, Bulgaria

P-96 Myocardial infarction and arterial thrombosis in homozygosity for the PAI-1 4G/5G polymorphism in identical newborn twins
De Lucia V. (1), Andreassi M.G. (2), Lunardini A. (1), Assanta N. (1), Spadoni I. (1), Giusti S. (2)
(1) Cardiologia Pediatrica, (2) Laboratorio di Biologia Cellulare e Genetica, CNR-Ospedale, G. Pasquinucci, Massa, Italy

P-97 The molecular genetic basis of complex congenital heart defects – a study of 47 families
Kwiatkowska J. (1), Wierzba J. (2), Aleszewicz-Baranowska J. (1), Erecinski J. (1).
Department of Paediatric Cardiology and Congenital Heart Disease, Medical University of Gdansk (1); Department of Paediatrics, Hematology, Oncology and Endocrynology, Medical University of Gdansk, Poland (2)

P-98 Tree classification analysis predicts prognosis at presentation in infantile i diopathic dilated cardiomyopathy
Azevedo V.M.P. (1), Santos M.A. (1), Castier M.B. (2), Amino J.G.C. (1), Cunha M.O.M. (1), Tura B.R. (1), Albanesi Filho F.M. (2), Xavier R.M.A. (1)
National Institute of Cardiology, Rio de Janeiro, Brazil (1); University of State of Rio de Janeiro, Rio de Janeiro, Brazil (2)

P-99 Predictors of supraventricular arrhythmia in adult patients with congenital heart diseases
Trojnarska O., Grajek S. (1), Kramer L. (2), Lanocha M. (1), Katarzynska A. (1)
1. University of Medical Sciencec Departmant of Cardiology,
2. University of Medical Sciences, Department of Computer Sciences and Staistic., Poznan, Poland

P-100 Electrocardiographic Features of Secundum Atrial Septal Defects with Pulmonary Hypertension in Adults
Ko J.K., Jhang W.K., Kim Y.H., Park I.S.
Department of Pediatrics, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea

P-101 Echocardiogram and equilibrium radionuclide ventriculography in the evaluation of right ventricular dysfunction in patients with transposition of great arteries and Mustard procedure
Marcora S., De Zorzi A., Garganese M., Cifra B., Silvetti M.S., Drago F., Giannico S.
Ospedale Pediatrico Bambino Gesù, Rome, Italy

P-102 Evaluation of sinus node function in young adults long term follow-up correction of d-transposition of great arteries by Senning atrial switch
Pietrucha A. Z. (1), Węgrzynowska M. (1), Pietrucha B.J. (2), Mroczek-Czernecka D. (1), Rudziński A. (2) Piwowarska W. (1)
Coronary Disease Department, Institute of Cardiology (1), Children Cardiology Department, Children University Hospital (2), Medical School of Jagiellonian University, Cracow, Poland

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P-103 Physical activity in children operated for congenital heart disease
Arvidsson A. (1), Slinde F. (1), Sunnegårdh J. (2), Hulthén L. (1)
Department of Clinical Nutrition, Sahlgrenska Academy at Göteborg University, Göteborg, Sweden (1); Queen Silvia Children´s Hospital, Sahlgrenska University Hospital, Göteborg, Sweden (2)

P-104 Neurodevelopmental and neuroradiologic outcome of UVH patients at age 5 to 7 years related to risk factor analysis
Sarajuuri, Anne, Jokinen E., Puosi R., Eronen M., Mildh L., Mattila I., Valanne L., Lönnqvist T.
Helsinki University Central Hospital, Hospital for Children and Adolescents, Helsinki, Finland

P-105 Natriuretic peptides within adults with secundum atrial septal defect (ASD II)
Lusawa T., Konka M., Janas J., Klisiewicz A., Demkow M., Hoffman P.
Institute of Cardiology, Warsaw, Poland

P-106 Patients with aortic coartation seen at a GUCH outpatient clinic
Urchaga A., Casaldàliga J., Girona J., Betrian P., Gran F., Manso B., Ferrer Q.
Unidad de Cardiopatias Congénitas del Adolescente y Adulto, Hospital Vall d'Hebron, Universitat Autònoma de Barcelona, Barcelona, Spain

P-107 Carotid-Subclavian Artery Index: Validation of an Echocardiographic Index to detect Coarctation
Mivelaz Y., Di Bernardo S., Meijboom E.J., Fall A.L., Sekarski N.
Pediatric Cardiology, Cardiovascular and Metabolic diseases Center, Lausanne Children's University Hospital (CHUV), Switzerland

P-108 Quantitative Assessment of Right Ventricular Function Using Three Dimensional SSFP Magnetic Resonance Angiography
Greil G.F. (1), Boettger Th. (2), Germann S. (3), Klumpp B. (4), Baltes Ch. (5), Kozerke S. (5), Bialkowski A. (3), Urschitz M.S. (6), Miller S. (4), Wolf I. (2), Hans-Peter Meinzer H.-P. (2), Sieverding L. (3)
Div. of Diagnostic Imaging and Dept. of Paediatric Cardiology, King´s College and Guy´s and St. Thomas´s Hospitals London, UK (1); Dept. of Medical and Biological Informatics, German Cancer Research Center, Heidelberg, Germany (2); Dept. of Pediatric Card

P-109 Does cardiac magnetic resonance imaging really replace diagnostic catheterization in congenital heart disease?
Sarikouch S., Schaeffler R., Haas N.A., Beerbaum P., Kececioglu D.
Department for Congenital Heart Disease, Heart and Diabetes-Center Northrine-Westfalia, Ruhr-University of Bochum, Bad Oeynhausen, Germany

P-110 Non-invasive Assessment of Congenital Pulmonary Vein Stenosis in Children Using Cardiac-non gated CT with 64-slice Technology
Marini D. (1), Agnoletti G. (1), Calcagni G. (1), Vouhé P. (1), Brunelle F. (2), Sidi D. (1), Bonnet D. (1), Ou P. (2)
Department of Pediatric Cardiology, Necker-Enfants Malades, Paris, France (1); Department of Pediatric Radiology, Necker-Enfants Malades, Paris, France (2)

P-111 Mitral Valve Annular Function in Children with Normal and Regurgitant Valves: A Three Dimensional Echocardiographic Study
Bharucha T., Sivaprakasam M.C., Roman K.S., Veldtman G.R., Vettukattil J.J.
Southampton Congenital Cardiac Centre, Southampton, UK

P-112 Intraoperative evaluation of a MicroMultiplane Transesophageal Echocardiographic Probe in Surgery for Congenital Heart Disease
Ten Harkel A.D.J. (1), Scohy T. (2), Gommers D. (2), Deryck Y. (2), McGhie J. (2), Bogers A.J.J.C. (2)
Erasmus MC-Sophia, Rotterdam, The Netherlands (1), Erasmus MC-Thoraxcenter, Rotterdam, The Netherlands (2)

P-113 Assessment of coronary artery aneurysms in patients with Kawasaki disease by multislice spiral computed tomography in follow-up
Kowalczyk M., Kosciesza A., Kawalec W., Turska-Kmiec; A., Ziolkowska L., Tomyn-Drabik M.
Children's Memorial Health Institute, Warsaw, Poland

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P-114 Tissue Doppler imaging analysis of right ventricular function before and after transcatheter closure of atrial septal defect
Laskari C., Kiaffas M., Tsutsinos A., Kantzis M., Papagiannis J., Rammos S.
Onassis Cardiac Surgery Center, Athens, Greece

P-115 QT and QTc intervals dispersion in children with chronic renal failure
Dimitriu A.G. (1), Hiastru G. (1), Brumariu O. (2), Munteanu M. (2), Pavel L. (1)
1. Ist Clinic of Pediatrics, University of Medicine and Pharmacy Iasi Romania;
2. IVth Clinic of Pediatrics,University of Medicine and Pharmacy Iasi Romania

P-116 Increased torsion, measured by Magnetic-Resonance-Tagging, of both ventricles in patients with Atrial Septal Defects normalises after interventional occlusion of the defect
Fratz S. (1), Keithahn A. (2), Lüchinger R. (3), Schwaiger M. (2), Hess J. (1), Stern H. (1)
Deutsches Herzzentrum München an der TUM, Munich, Germany (1); Klinikum rechts der Isar an der TUM, Munich, Germany (2); University and Swiss Federal Institute of Technology, Zurich, Switzerland (3)

P-117 Interventional treatment in postoperative pulmonary stenosis and hypoplasia in children with HLHS during multistage operative treatment
Moszura T. (1), Mazurek-Kula A. (1), Dryzek P. (1), Moll J.A. (1), Moll J.J. (2), Sysa A. (1)
Cardiology Department Polish Mother's Memorial Hospital, Research Institute (1), Cardiosurgery Department Polish Mother's Memorial Hospital, Research Institute (2) Lodz, Poland

P-118 Transcatheter Recanalization of Acutely Occluded Aortopulmonary Shunts
Sreeram N., Emmel M., Brockmeier K., Bennink G.
University Hospital of Cologne, Cologne, Germany

P-119 Stenting of the transverse arch in arch hypoplasia after surgical repair of coarctation of the aorta
Haas N.A., Schaeffler R., Laser T., Beerbaum P., Wegendt C., Goerg R., Sarikouch S., Matthies W., Kececioglu D.
Heart and Diabetes Centre North-Rhine Westfalia, Bad Oeynhausen, Germany

P-120 Dilatation of aortic coarctation in infantswith severe left ventricular dysfunction: a bridge to surgery
Bouzguenda I., Marini D., Boudjemline Y., Bonnet D., Agnoletti G.
Necker Enfants Malades, Paris, France

P-121 Pulmonary stent implantation in children with single ventricle before and after completion of cavopulmonary connection
Kretschmar O., Knirsch W. (1), Balmer C. (1), Berger F. (3), Prêtre R. (2)
University Children`s Hospital, Department of Pediatric Cardiology (1) and Department of Cardiothoracic Surgery (2), Zurich, Switzerland; German Heart Center Berlin, Department of Pediatric Cardiology (3), Berlin, Germany

P-122 Mid-long term follow-up results of stent dilatation of Aortic coarctation
Ait-Ali L., Spadoni I., Assanta N., Festa P., Carminati M., Carducci T., Giusti S.
Pediatric Cardiology and GUCH Unit, CNR, "G. Pasquinucci" Hospital, Massa, Italy

P-123 Transcatheter Duct Occlusion in Adult Patients
Anjos R., Ferreira R., Bento A., Teixeira A., Rossi R., Menezes I., Maymone-Martins F.
Hospital de Santa Cruz, Lisbon, Portugal

P-124 Self-expanding nitinol stents may improve Right-ventricular-to-Pulmonary artery-Conduit obstructions after Norwood-Operation
Gitter R. (1), Mair R. (2), Lechner E. (1), Tulzer G. (1)
Department of Pediatric Cardiology, Childrens Hospital Linz, Austria (1); Department of Cardio-Thoracic-Surgery, General Hospital Linz, Austria

P-125 Interventional management of pulmonary sequestration
Witsenburg M., De Jong P.L., Dalinghaus M., Du Plessis F.
ErasmusMC-Sophia Children's Hospital Rotterdam Netherlands

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P-126 **Safety and efficacy of different minimally invasive atrial septal defect closure**
Colaneri M.°, Quarti A. , D' Alfonso A.*, Baldinelli A.°, Ricciotti R.°, Bettuzzi M.G.°, Munch C.^, Pozzi M.**
*°Paediatric Cardiology – *Paediatric Surgery – ^SOD Anestesia Rianimazione Presidio Cardiologico "G.M. Lancisi" – Ospedali Riuniti – Ancona – Italy*

P-127 **Long term results of Fontan operation: twenty years clinical experience at a single medium volume center**
*Stellin G. (1), Padalino M.A. (1), Lo Rito M. (1), Milanesi O. (2), Biffanti R. (2)
Pediatric and Congenital Cardiovascular Surgery (1), Pediatric Cardiology (2)*

P-128 **Integrating post operative value of cardiac Troponin-I for a better correlation with in-hospital outcomes after congenital heart surgery**
*Di Bernardo S. (1), Stucki P. (2), Perez M.-H., Racine L. (2), Hurni M. (3), Mivelaz Y. (1), Bernath M.-A. (4), Cotting J. (2), Sekarski N. (1)
Pediatrics Cardiology, University Hospital, Lausanne, Switzerland (1); PICU, University Hospital, Lausanne, Switzerland (2), cardio-vascular surgery, University Hospital, Lausanne, Switzerland (3); Pediatric anaesthesiology, University Hospital, Lausanne*

P-129 **Acute Neurologic Complications Occurred After Cardiac Surgery In Children**
*Piazza L. (1)., Micheletti A. (1), Allegri V. (2), Sansone V. (1), Negura D. (1), Butera G. (1), Chessa M. (1), Arcidiacono C. (1), Rosti L. (1), Fontana A. (1), Cotticelli B. (1), Meola G. (1), Frigiola A. (1), Carminati M. (1)
Dipartimento di Cardiologia e Cardiochirurgia Pediatrica, IRCCS Policlinico S.Donato, S.Donato Milanese (MI), Italy (1); Dipartimento di Neurologia, Università degli Studi di Milano, IRCCS Policlinico S.Donato, S.Donato Milanese (MI) Italy, (1); Unità Op*

P-130 **Conversion of Atrio-pulmonary Connection to Total Cavo-pulmonary Connection: A Single Centre Experience with the Fontan Circulation**
*Greutmann M. (1), Trigo Trindade P. (1), Kretschmar O. (2), Dodge-Khatami A. (3), Prêtre R. (3), Bauersfeld U. (2)
Cardiology, University Hospital Zurich, Switzerland (1); Pediatric Cardiology, University Children's Hospital Zurich, Switzerland (2); Clinic for Cardiovascular surgery, University Children's Hospital, Zurich, Switzerland (3)*

P-131 **Initial experience with unifocalization and repair of pulmonary atresia with VSD and major aortopulmonary collaterals in Lund**
*Johansson J. (1), Hanseus K. (2), Jögi P. (1), Malm T. (1), Rehnström P. (1), Ville J. (2), Johansson S. (1)
Department of Paediatric Cardiac Surgery (1), Department of Paediatric Cardiology (2), Lund, Sweden*

P-132 **Extra-anatomic conduits for treatment of adult hypoplasia or interruption of the aortic arch**
*Marques M., Abecasis M., Rodrigues R., Boshoff S., Hernandez R., Figueiredo S., Anjos R., Melo J.
Hospital de Santa Cruz, Lisbon, Portugal*

P-133 **Predictors of Preoperative Neurological Outcome and its Impact on Surgery in Children with Congenital Heart Disease undergoing Cardiopulmonary Bypass**
*Lianos A. (1), Waldvogel K. (2), Molinari L. (1), Dodge-Khatami A. (3), Kaufmann M. (1), Bauersfeld U. (4), Latal B. (1)
Child Development Center (1), Intensive Care Unit (2), Division of Congenital Cardiovascular Surgery (3), Division of Cardiology (4), University Children's Hospital, Zurich, Switzerland*

20.00 **Gala dinner**

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Saturday, 19.05.2007

08:00–09:00 **AEPC Working Groups Business Meetings**

Basic Genetics Working Group	Aula A
Morphology Working Group	Aula B
Council Meeting – part I	Hall 1

09:00–10:30 **Basic Genetics of Congenital Heart Defects Working Group** Aula A

Advances in the understanding of genetics and basic sciences in valvar congenital heart disease
Chairs: J. Parsons (Leeds, UK), M. Krajewska-Walasek (Warsaw, PL)

The genetics of valvular malformations
G. Andelfinger (Montreal, CAN)

Myxoid degeneration of the atrioventricular valves – New Genetic insights
P. Bouvagnet (Lyon, FR)

Advances in tissue engineering in the development of new heterograft valves
E. Ingham (Leeds, UK)

09:00–10:30 **Morphology Working Group Session** Aula B

Update on cardiomyopathies
Chairs: A. Angelini (Padua, IT), W. Kawalec (Warsaw, PL)

Contemporary classification on cardiomyopathies
G. Thiene (Padua, IT)

Arrhythmogenic right ventricular dysplasia/cardiomyopathies
E. K. Wlodarska (Warsaw, PL)

Non compation myocardium
C. H. Attenhofer Jost (Zurich, CH)

10:40–11:40 **Abstract Session 10: General Paediatric Cardiology** Aula A
Chairs: E. Jokinen (Helsinki, FL), J. Bialkowski (Zabrze, PL)

10:40 **O10-1** **Fetal echocardiography in the first trimester for fetuses at risk of congenital heart disease**
Lopes K. (1), Iserin F. (1), Oury J.F. (2), Platet A. (2), Armoogum P. (3), Azancot A. (1)
Fetal cardiac unit (1), obstetrics (2), INSERM CIE5 (3), Hopital Robert Debre Paris, France

10:50 **O10-2** **Fetal left heart obstructions-diagnosis, development and outcome during the first year of life**
Oberhoffer R. (1,2), Zimmermann A. (1), Czettritz M. (1), Schneider K.T.M. (1), Hess J. (2)
Fetomaternal Center Technical University Munich (1), German Heart Center Munich (2)

11:00 **O10-3** **Results of stent implantation in 7 neonates with pulmonary atresia and left pulmonary artery stenosis via modified Blalock-Taussig shunts**
Zubrzycka M., Brzezinska-Rajszys G., Kansy A., Maruszewski B., Rewers B., Ksiazyk J., Turska-Kmiec A., Daszkowska J.
The Children Memorial Health Institute, Warsaw, Poland

11:10 **O10-4** **Catheterinterventional treatment of Sano shunt obstruction**
Daehnert I., Riede F.T., Razek V., Weidenbach M., Rastan A., Walther T., Kostelka M.
Herzzentrum, University of Leipzig, Germany

11:20 **O10-5** **Aortic Dilation and Rearrangement of the Aortic Root in Turner Syndrome: Comparative Assessment Using Transthoracic Echocardiography and Magnetic Resonance Imaging**
Prandstraller D. (1), Giardini A. (1), Mazzanti L. (2), Sciarra F. (1), Lovato L. (3), Fattori R. (3), Cicognani A. (2), Picchio F.M. (1)

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Pediatric Cardiology and Adult Congenital Unit, University of Bologna, Italy (1); Department of Pediatrics, University of Bologna, Italy (2); Cardiac MRI Unit, University of Bologna, Italy (3)

11:30 **O10-6** **Two years follow-up of the "growth stent" – results in infants with aortic coarctation**
Ewert P. (1), Peters B. (1), Miera O. (1), Nagdyman N. (1), Ovroutski S. (1), Stiller B. (1), Schulze-Neick I. (1), Berger F. (2)
Dept. of Congenital Heart Disease, German Heart Institute Berlin, Berlin Germany (1); Klinik für Paediatrie mit Schwerpunkt Kardiologie, Universitätsklinikum Charité, Berlin, Germany (2)

10:40–11:40 **Abstract Session 11: Intervention** Aula B
Chairs: J. Butera (Milano, IT), M. Witsenburg (Rotterdam, NL)

10:40 **O11-1** **From complications to changes in approach – experience with percutaneous pulmonary valve implantation**
Nordmeyer J. (1), Lurz P. (1), Bolger A.P. (2), Coats L. (1), Frigiola A. (1), Walker F. (2), Cullen S. (1,2), Bonhoeffer P. (1,2).
UCL Institute of Child Health and Great Ormond Street Hospital for Children, London, UK (1); The Heart Hospital, London, UK (2)

10:50 **O11-2** **A new hybrid therapy for closure of muscular ventricular septal defects in a pig model**
Kozlik-Feldmann R. (1), Lang N. (1), Aumann R. (1), Sodian R. (2), Rassoulian D. (2), Freudenthal F. (5), Hinterseer M. (3), Daebritz S. (2), Netz H. (1), Vasilyev N. (4), Del Nido P. (4)
Pediatric Cardiology, Munich, Germany (1); Cardiac Surgery, Munich, Germany (2); Medicine I,Munich, Germany (3); Cardiac Surgery, Boston, USA (4); Kardiozentrum, La Paz, Bolivia (5)

11:00 **O11-3** **Early experience with dilatable pulmonary artery band and interventional device closure of muscular ventricular septal defects**
Yates R., Gnanakanthan K., Kanani M., Bonhoeffer P., Tsang V.
Great Ormond Street Hospital NHS Trust, London, UK

11:10 **O11-4** **Real-time Left Ventricular Physiology During Transcatheter Closure of Atrial Septal Defects Measured by Conductance Catheter**
Lunze K., Ewert P., Peters B., Miera O., Berger F., Schulze-Neick I.
German Heart Institute Berlin, Germany

11:10 **O11-5** **Stent Implantation for the Treatment of Adult Aortic Coarctation: Initial and Five-Year Results**
Thanopoulos B.D. (1), Basta E. (1), Loukopoulou S. (1), Eleptherakis N. (1), Paphitis CH. (1), Skoularigis I. (2), Tryposkiadis F. (2), Zarayelyan A. (3)
"Aghia Sophia" Children's Hospital, Athens, Greece, [1] University Hospital of Thessaly, Larissa,Greece, [2] Yerevan State Medical University Hospital, Yerevan, Armenia [3]

11:20 **O11-6** **Stent implantation in aortic coarctation: Bare or Covered?**
Butera G., Piazza L., Chessa M., Abella R., Micheletti A., Negura D., Arcidiacono C., Rosti L., Fesslova V., Carminati M.
Pediatric Cardiology – Policlinico San Donato IRCCS – Italy

10:40–11:40 **Abstract Session 12: Basic Science** Hall 1
Chairs: J. Parsons (Leeds, UK), P. Bouvagnet (Lyon, FR)

10:40 **O12-1** **Presence of accessory pathways in the developing human heart, possible explanation for fetal and neonatal atrioventricular reentrant tachycardias**
Hahurij N.D. (1), Blom N.A. (1), Kolditz D.P. (2), Wijffels M.C.E.F. (2), Bökenkamp R. (1), Markwald R.R. (3), Schalij M.J. (2), Poelmann R.E. (4), Gittenberger-de Groot A.C. (4)
Department of Paediatric Cardiology, Leiden University Medical Center, Leiden, The Netherlands (1); Department of Cardiology, Leiden University Medical Center, Leiden, The Netherlands (2); Department of Cell Biology and Anatomy, Medical University of Sout

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10:50 **O12-2** **Impact of the matrix used in tissue-engineered heart valves on bacterial adhesion in a model of endocarditis**
Heying R. (1), Wolf C. (2), Schmidt K.G. (1), Schroten H. (2)
Department of Paediatric Cardiology (1); Paediatric Infectious Diseases, Department of General Paediatrics (2), University Hospital Duesseldorf, Germany

11:00 **O12-3** **Differential proteomic analysis of the pressure-overloaded right ventricle of young rats over time reveals specific changes in heat shock- and antioxidant proteins**
Faber M.J. (1), Dalinghaus M. (1), Lankhuizen I.M. (1), Bezstarosti K. (2), Duncker D.J. (3), Helbing W.A. (1), Lamers J.M.J. (2)
Erasmus MC – Sophia, Dept. Pediatrics, Div. Pediatric Cardiology, Rotterdam, The Netherlands (1); Erasmus MC, Dept. Biochemistry, Rotterdam, The Netherlands (2); Erasmus MC, Dept. Experimental Cardiology, Rotterdam, The Netherlands (3)

11:10 **O12-4** **Stem Cells recruitment in Surgical palliation for Hypoplastic Left Heart Syndrome**
Castellani C. (1), Padalino M.A. (2), della Barbera M. (1), Toffoli S. (1), Milanesi O. (3), Stellin G. (2), Thiene G. (1), Angelini A. (1).
Cardiovascular Pathology (1), Pediatric and Congenital Cardiovascular Surgery Unit (2), Pediatric Cardiology (3), University of Padua, Medical School, Italy

11:20 **O12-5** **Extracardiac Progenitor Cells Repopulation in Pediatric sex mismatch heart transplants**
Castellani C. (1), della Barbera M. (1), Tona F. (2), Caforio ALP. (2), Gambino A. (3), Valente M. (1), Gerosa G. (3), Thiene G. (1), Angelini A. (1)
Cardiovascular Pathology (1), Division of Cardiology (2), Dept. of Cardiovascular Surgery (3), University of Padua, Medical School, Italy

11:30 **O12-6** **Exercise training does not improve significantly endothelial function in pre pubertal obese children but prevent intima media thickening**
Aggoun Y., Golay E., Keller-Marchand L., Farpour-Lambert N.J., Beghetti M.
Pediatric Cardiology Unit, Geneva, Switzerland

11:40–12:15 **Coffee Break**

12:15–13:15 **State of the Art Session: Arrhythmia** Aula
Chairs: A. Celiker (Ankara, TR), K. Bieganowska (Warsaw, PL)

12:15–12:40 **Catheter ablation – lessons learnt**
T. Paul (DE)

12:40–13:05 **Resynchronization and prosynchronization- multifaceted/multidisciplinary approach**
J. Janousek (Leipzig, DE)

13.05–13.15 **Discussion**

13:30–14:30 **Abstract Session 13 General Paediatric Cardiology** Aula A
Chairs: A. Bozio (Lyon, FR), J.P. Pfammatter (Berne, CH)

13:30 **O13-1** **Prognostic Value of B-Type Natriuretic Peptide in Children with Pulmonary Hypertension**
Lammers A.E. (1), Hislop A.A. (2), Bonhoeffer P. (1), Haworth S.G. (1,2)
Great Ormond Street Hospital for Children, London, UK (1); Institute of Child Health, London, UK (2)

13:40 **O13-2** **Beneficial effects of ghrelin in a new model of infantile pulmonary hypertension induced by monocrotaline**
Neves A.L. (1,2), Santos M. (1), Henriques-Coelho T. (1,3), Oliveira S. M. (1,5), Baptista M.J. (2,4), Areias J.C. (2), Correia-Pinto J. (3,4), Leite-Moreira A. (1, 6)
Department of Physiology, Faculty of Medicine, Oporto, Portugal (1); Department of Paediatric Cardiology, Hospital of São João, Oporto, Portugal (2); Department of Paediatric Surgery, Hospital São João, Oporto, Portugal (3), Health and Life Sciences Resea

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13:50 **O13-3** **Parameters of heart rate variability predict outcome in children with pulmonary hypertension**
Lammers A.E. (1), Munnery L.(1), Hislop A.A. (2), Bonhoeffer P. (1), Haworth S.G. (1)
Great Ormond Street Hospital for Children,London, UK (1); Institute of Child Health, London, UK (2)

14:00 **O13-4** **The use of Bosentan in the treatment of Pulmonary Arterial Hypertension in infants less then one year of age**
Prendiville T., McMahon C., Oslizlok P.
Our Lady's Hospital for Sick Children, Ireland

14:10 **O13-5** **Cardiac Autonomic Nervous Activity is Impaired in Patients with the Eisenmenger Syndrome and Tends to Improve Under Treatment with Bosentan**
Lunze K. (1), Farahwaschy B. (1), v. Bismarck I. (1), Gilbert N. (1), Mebus S. (1), Wensel R. (2), Schlehofer H. (3), Opitz C. (3), Berger F. (1), Schulze-Neick I. (1)
German Heart Institute Berlin, Germany (1); Klinikum der Universität Regenburg, Germany (2); DRK Kliniken Berlin Westend, Germany (3)

14:20 **O13-6** **Early Predictors of Cardiomyopathy in Adolescents with Type I Diabetes**
El Behery S. (1), Monem A. (1), Khalil D. (1), El Hadidi E. (2), Salem M. (1)
From the Pediatric (1) and Clinical Pathology (2) Departments, Ain Shams University Faculty of Medicine, Cairo, Egypt

13:30–14:30 **Abstract Session 14: General Paediatric Cardiology** Aula B
Chairs: J. de Giovanni (Birmingham, UK), P. Burczynski (Warsaw, PL)

13:30 **O14-1** **Transcatheter ASD closure in children below 13 kg**
Szkutnik M. (1), Brzeziska Rajszys G. (2), Kusa J. (1), Zubrzycka M. (2), Banaszak P. (1), Ksiazyk J. (2), Rewers B. (1), Bialkowski J. (2)
Congenital Heart Disease and Pediatric Cardiology Dept., Silesian Center for Heart Diseases, Zabrze, Poland (1); Heart Catheterization Laboratory, The Children Memorial Health Institute, Warsaw, Poland (2)

13:40 **O14-2** **Results of transcatheter closure of unroofed coronary sinus**
Wang J.K., Tsai S.K., Chen S.J., Wu M.H.
Departments of Pediatrics, National Taiwan University Hospital, College of Medicine, National Taiwan University, Taipei, Taiwan

13:50 **O14-3** **Effectiveness of transcatheter patch release with surgical adhesives in the occlusion of heart defects**
Calachanis M., Macuil B., Zamora R., Coulson J., Toumanides S., Sideris E.
Athenian Institute of Pediatric Cardiology, Athens, Greece

14:00 **O14-4** **Use Of Stents In Patients With Congenital Heart Defects**
Alekyan B.G., Podzolkov V.P., Pursanov M.G., Gadghiev A.A., Zelenikin M.A., Shatalov K.V., Kokshenev I.V.
Bakoulev scientific Center for Cardiovascular Surgery RAMS, Moscow, Russia

14:10 **O14-5** **Use of covered stents in the management of native coarctation of the aorta-Diversiry of indications and a special place in developing countries**
Sadiq M., Rehman A., Hyder N., Younis M., Masud F., Wilkinson J.W., Qureshi S.A.
Punjab Institute of Cardiology and The Children's Hospital, Lahore, Pakistan

14:20 **O14-6** **Promotion of Growth of True Pulmonary Arteries as a First Step in Staged Repair of Pulmonary Atresia, Ventricular Septal Defect and Major Aorto-Pulmonary Collateral Arteries**
Rammeloo L., Hruda J., Blom N.A., Bökenkamp R., Ottenkamp J., Hazekamp M.G.
Center for Congenital Heart Defects Amsterdam-Leiden, the Netherlands

13:30–14:30 **Abstract Session 15: General Paediatric Cardiology** Hall 1
Chairs: J. Weil (Hamburg, DE), F. Godart (Lille, FR)

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13:30 **O15-1** **Persistent Ductus Arteriosus – more than an isolated anomaly**
deBruijn L. (1), Houwing-Duistermaat J.J. (2), Tanke S. (1), Hruda J. (1), Ottenkamp J. (1), Bokenkamp R. (1)
(1) Center of Congenital Anomalies of the Heart Amsterdam/Leiden (CAHAL) and (2) Department of Medical Statistics Leiden University Medical Center, Leiden, NL

13:40 **O15-2** **Determination of biventricular diastolic compliance and myocardial contractility using Real-Time MRI: a In-Vivo Validation Study**
Kuehne T., Lunze K., Maarouf N., Rahmanzadeh P.*, Pietzner K., Meinus C., Krüger J., Schulze-Neick I., Berger F.*
Deutsches Herzzentrum Berlin, Department of Congenital Heart Diseases and Pediatric Cardiology

13:50 **O15-3** **Cardiovascular Findings of Tricyclic Antidepressant Intoxication In Children**
Dinleyici E.C. (1), Kilic Z. (2), Aydin B. (1), Yarar C. (1), Yildiz B. (1), Ucar B. (2)
Department of Pediatrics (1) and Pediatric Cardiology (2), Eskisehir Osmangazi University Faculty of Medicine, Eskisehir, Turkey

14:00 **O15-4** **Integration of in silico analyses from small pharmacokinetic studies into randomized controlled trials avoid confounding by dose selection in paediatric drug therapy**
Albers S. (1), Meibohm B. (2), Barrett J. (3), Mir T.S. (4), Laer S. (1)
Clinical Pharmacy and Pharmacotherapy, University of Düsseldorf, Germany (1), Department of Pharmaceutical Sciences, University of Tennessee, Memphis, USA (2), Childrens Hospital of Philadelphia, Phildelphia, USA (3), Paediatric Cardiology, University of Hamburg, Germany (4)

14:10 **O15-5** **Telemedicine as discharge support for families of children with major congenital heart disease**
McCrossan B., Morgan G., Grant B., Sands A., Craig B. & Casey F.
Department of Paediatric Cardiology, Royal Belfast Hospital for Sick Children, UK

14:20 **O15-6** **Effects of a 3-month exercise intervention on ambulatory blood pressure and cardiovascular disease risk factors in prepubertal obese children**
Farpour-Lambert N.J. (1), Aggoun Y. (1), Keller-Marchand L. (1), Schwitzgebel V. (2), Herrmann F.R. (3), Beghetti M.(1)
Pediatric Cardiology Unit, Dept of Pediatrics (1); Pediatric Endocrinology and Diabetology Unit, Dept of Pediatrics (2); Geriatrics Division, Dept of Rehabilitation and Geriatrics (3); Geneva University Hospitals, Switzerland

14:40 **AEPC Council Meeting part II** Hall 1

15:00 **Post Congress Tour**

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Cardiology in the Young

42nd Annual Meeting
Association for European Paediatric Cardiology, 16–19 May, 2007; Warsaw Medical Academy Congress Center

O1-1

The Impact of Multiplanar Review of 3-Dimensional Echocardiographic Data on Management of Congenital Heart Disease

Bharucha T., Roman K.S., Vettukattil J.J.
Congenital Cardiac Centre, Southampton, UK

Introduction: Accurate diagnosis of functional anatomy is of critical importance to management decisions.
The multiplanar review mode (MPR) allows the operator to view the moving 3D data set in 3 orthogonal planes simultaneously, and to review the image in infinite planes. We sought to investigate the feasibility and clinical utility of MPR in congenital heart disease, and to compare MPR findings to 2D echocardiography.
Methods: Between October 2004 and December 2006, 2 and 3-dimensional data sets were acquired in 300 consecutive patients, all unsedated and spontaneously breathing. Two-dimensional findings were analysed and recorded, and MPR analysis performed subsequently and additional findings recorded.
Results: The main diagnoses were as follows: Valve abnormalities 106 (35.3%), subaortic stenosis 18 (6%), left-to-right shunts 46 (15.3%), complex 95 (31.7%) (AVSD, DORV, HLHS, tetralogy of Fallot, TGA, tricuspid atresia, truncus arteriosus), anomalous pulmonary venous connections 4 (1.3%), aortic arch anomalies 7 (2.3%), hypertrophic cardiomyopathy 2 (0.7%), normal 22 (7.3%). MPR analysis was possible in all patients.
In 33 (11%) cases, management was substantially altered by new MPR-derived information as follows:

- 16 - Atrioventricular valve morphology; facilitation of corrective surgery in 13 of these, unnecessary surgery prevented in 3.
- 8 - Ventricular size and morphology, altering decision to pursue single ventricle palliation or biventricular repair.
- 5 - Mechanism of aortic valve stenosis or regurgitation.
- 4 - Great vessel relationships.
- 2 - Mechanism of para-prosthetic valve leak.
- 1 - Vascular ring demonstrated, without requirement for further imaging.

Conclusions: 3D MPR is feasible in congenital heart disease, allowing analysis despite poor resolution with 3D volume-rendering.
MPR provides additional information to 2D echocardiography that substantially alters clinical management in many patients with complex congenital heart disease.
We recommend the use of 3D MPR for patients with complex atrioventricular valves or ventricular morphology, and great vessel relationships.

O1-2

Evidence of liver fibrosis in children with Fontan circulation. Noninvasive assessement by a novel Doppler elastography and biochemical marker test

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Basics: Mid term studies have shown, that patients who received the Fontan procedure can develop liver cirrhosis with its sequelae. Therefore, early staging of liver fibrosis is essential to detect patients at risk for developing liver cirrhosis. Transient elastography (FibroScan) has been intensively evaluated for non-invasive staging of liver fibrosis in adults with good results. The best results for the noninvasive staging of liver fibrosis were achieved by the combination of FibroScan and serum fibrosis markers. Here, we investigated the liver fibrosis stages in patients following the Fontan procedure using the FibroScan and FibroTest.
Methods: 39 patients following the Fontan procedure were included in the study. Patients baseline characteristics were determined by age (11.55 years+/–5.41), time interval since the Fontan operation (5.647 years+/–3.44). All patients received an ultrasound of the liver and spleen, including Duplex and Doppler ultrasound and the transient elastography (FibroScan). In addition, detailed laboratory testing was performed including Apoprotein A1, α2-Macroglobulin and Haptoglobin for calculation of the FibroTest and ActiTest.
Results: The Spearman correlation coefficient between the liver stiffness measured with the FibroScan and the time interval since

the Fontan operation was highly significant with R = 0.514 (p = 0.001). No significant correlation could be found between the fibrosis staging using the FibroTest and the time interval since the Fontan operation (R = 0.225, p = 0.201). However, significant correlation was found between the FibroTest values and the age of the patient (R = 0.479, p < 0.005). In addition a significant correlation was found between the ActiTest calculated from the liver biochemical markers and the time interval since the Fontan procedure (0.374, p < 0.05).
Discussion: The present study shows that patients following the Fontan procedure are at increased risk to develop liver fibrosis and liver cirrhosis. The risk increases with the age of the patient and the time interval since the Fontan procedure. The non-invasive measurement of liver fibrosis using the FibroScan and FibroTest can be a useful tool to identify Fontan patients at risk.

O1-3
Assessment of pulmonary artery size and function with phase contrast magnetic resonance imaging in patients after Fontan operation at young age

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Introduction: Little is known about the effects of long-term non-pulsatile flow on pulmonary artery (PA) growth after Fontan operation. Furthermore, the effects on flow variables and shear stress have not been studied in a patient group operated on at young age. In this study, we assessed PA size, flow variables, and shear stress long-term after Fontan operation at young age and compared them with healthy controls, using cardiovascular magnetic resonance (CMR) imaging.
Methods: 14 patients (9 males, aged 13.1 ± 4.0 years, follow-up after Fontan completion 9.7 (5.4–16.8) years) and 17 healthy controls 9 males, aged 13.3 ± 2.3 years) were included. Flow measurements in the branch PA were made during a CMR study, using phase contrast velocity-encoded imaging. In patients, flow measurements were repeated during low-dose dobutamine stress of 7.5 μg/kg/min. Shear stress was determined according to a previously published method (J Am Coll Cardiol 2005;45:846–854).
Results: CMR scanning and dobutamine administration was well tolerated by all subjects without side effects. Results of the flow studies and shear stress determination are summarized in the following table (NS = not significant):

	controls	patients, rest	patients, stress	p-value (controls vs patients)	p-value (rest vs stress)
Heart rate (/min)	72 ± 12	69 ± 12	93 ± 17	NS	<0.001
Stroke index (ml/m^2)	31 ± 7	19 ± 7	19 ± 7	<0.001	NS
Total flow (ml/min/m^2)	2189 ± 463	1244 ± 74	1705 ± 308	<0.001	<0.001
Average flow (ml/s)	56 ± 15	28 ± 6	39 ± 13	<0.001	<0.001
Peak flow (ml/s)	187 ± 48	55 ± 31	71 ± 44	<0.001	<0.001
Diameter (mm)	16.2 ± 1.7	15.1 ± 2.7	15.2 ± 2.8	NS	NS
Distensibility	0.41 ± 0.09	0.22 ± 0.06	0.20 ± 0.07	<0.001	NS
Shear stress (N/m^2)	0.84 ± 0.14	0.38 ± 0.15	0.50 ± 0.18	<0.001	<0.001

Conclusions: PA diameter is normal in patients long-term after Fontan operation at young age. However, flow variables, distensibility and shear stress are significantly lower compared to healthy controls, implicating pulmonary endothelial and/or vascular dysfunction.

O1-4
Aortic root dysfunctioning and its effect on left ventricular function after the arterial switch operation assessed with MRI

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Objectives: Evaluation of aortic wall elasticity, aortic dimensions, aortic valve competence and left ventricular (LV) function in patients late after the arterial switch operation (ASO) with magnetic resonance imaging (MRI).
Background: Intrinsic pathology of the aortic wall is a possible explanation for aortic dilatation in patients late after ASO. The relationship between aortic wall elasticity, aortic dimensions, aortic valve competence and LV function in ASO patients has not previously been studied.
Methods: MRI was performed in 15 ASO patients (mean ± SD age (yrs.): 16 ± 4) and 15 matched controls.
Results: Reduced aortic elasticity in ASO patients was indicated by increased pulse wave velocity (PWV) in the aortic arch (5.1 m/s ± 1.2 vs. 3.9m/s ± 0.7, P = 0.004) and reduced root distensibility ($2.2\star10^{-3}$ mmHg^{-1} ± 1.8 vs. $4.9\star10^{-3}$ mmHg^{-1} ± 2.9, P < 0.01). ASO patients frequently showed aortic root dilatation as compared to controls (mean difference 5.7–9.4 mm, P ≤ 0.007 at 3 predefined levels). Minor degrees of aortic regurgitation (AR) were present in 6 patients (AR fraction 5% ± 3 vs. 1% ± 1, P < 0.001). In addition, LV ejection fraction (LV EF) was decreased in ASO patients (51% ± 6 vs. 58% ± 5, P = 0.003) and LV dimensions were enlarged with LV end-diastolic volume 112 mL/m^2 ± 13 vs. 95 mL/m^2 ± 16, P = 0.007 and LV end-systolic volume 54 mL/m^2 ± 11 vs. 39 mL/m^2 ± 7, P < 0.001. AR fraction was correlated with increased PWV in the aortic arch (r = 0.39, P = 0.03), reduced root distensibility (r = 0.45, P = 0.01) and aortic root dilatation (r 0.32–0.66, P ≤ 0.01 for all levels). Degree of AR predicted decreased LV EF (r = 0.41, P = 0.026) and was correlated with increased LV dimensions (r = 0.48, P = 0.008; r = 0.67, P < 0.001; respectively).
Conclusion: Reduced elasticity of the proximal aorta and aortic root dilatation were frequently present in ASO patients. Dilatation of the aortic root and reduced proximal aortic wall elasticity were associated with degree of AR, while AR was correlated with reduced LV systolic function and increased LV dimensions late after ASO.

O1-5
Noncompaction of the left ventricular myocardium in children: echocardiographic response to therapy

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Introduction: Noncompaction of the left ventricular myocardium was included in the 2006 classification of cardiomyopathies as a Genetic Cardiomyopathy and occurs due to a disorder of endomyocardial morphogenesis resulting in a failure of trabecular compaction of the developing myocardium.
Objectives: The aim of the present study was to describe the echocardiographic follow up of ventricular noncompaction in paediatric patients treated for heart failure.

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Methods: Seventeen patients (8 males and 9 females), from 1 month to 14 years were enrolled in a longitudinal cohort study for a mean peirod of 3.3+/−4.5 years (1 month to 14 years). Serial echocardiographic examinations were performed to assess systolic and diastolic function.
Results: Left ventricular ejection fraction (EF) at diagnosis was 36.4+/−14.2% and improved to 52.2+/−19.3% at last follow up ($p < 0.000$). Only four patients had a EF above 40% at diagnosis. Diastolic pattern was restrictive at diagnosis in 58.8% of the patients, whereas at follow up only 41.2% persisted with a restrictive pattern. Restrictive pattern was not present when EF was above 40%. Twelve patients were treated with carvedilol on top of the conventional heart failure therapy and this group showed an improvement of the EF from 36.1+/−10.7% to 44.2+/−15.1% ($p < 0.000$). Two patients died and one is being listed for heart transplantation. One patient had a neurological thromboembolic event.
Conclusions: Noncompaction of the left ventricular myocardium has a wide spectrum of clinical and echocardiographic presentation in children. They seem to improve with conventional heart failure therapy with a better response to carvedilol. A greater number of patients must be studied in a randomized fashion in order to provide more accurate answers to this entity.

O1-6
MRI of aortic wall elasticity, aortic valve competence and LV function in patients with a non-stenotic bicuspid aortic valve
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Introduction: Intrinsic pathology of the aortic wall is a possible explanation for aortic dilatation in patients with a bicuspid aortic valve (BAV), even in the absence of a stenotic aortic valve. The relationship between aortic dimensions, aortic wall elasticity, aortic valve competence and left ventricular (LV) function in patients with BAVs has not previously been studied.
Purpose: To evaluate aortic dimensions, aortic wall elasticity, aortic valve competence and LV function in patients with a non-stenotic BAV using magnetic resonance imaging (MRI).
Methods: MRI was performed in 20 patients with non-stenotic BAVs (13 male; mean ± SD age (yrs.): 27 ± 11) and 20 matched healthy subjects. Aortic root diameters at 4 predefined levels, aortic valve competence and systolic LV function were measured using standard MRI sequences. Pulse wave velocity (PWV) in the aortic arch and descending aorta, and aortic root distensibility were used as markers of aortic wall elasticity.
Results: Patients with BAVs showed aortic root dilatation as compared to controls (mean difference 3.6–4.2mm, with $P \leq 0.04$ at all 4 levels). Increased PWV in the aortic arch and descending aorta was observed in patients (5.6m/s ± 1.3 vs. 4.5m/s ± 1.1, $P = 0.01$; and 5.2m/s ± 1.8 vs. 4.3m/s ± 0.9, $P = 0.03$, respectively), as well as reduced aortic root distensibility ($3.1 \star 10^{-3}$ mmHg^{-1} ± 1.2 vs. $5.6 \star 10^{-3}$ mmHg^{-1} ± 3.2, $P < 0.01$). Minor degrees of aortic regurgitation (AR) ranging between 5 and 16% were present in 11 patients (AR fraction 6% ± 8 vs. 1% ± 1, $P < 0.01$). LV ejection fraction was normal (55% ± 8 vs. 56% ± 6, $P = 0.61$), whereas LV mass was significantly increased in patients (54 gr/m^2 ± 12 vs. 46 gr/m^2 ± 12, $P = 0.04$). Aortic root dilatation (r ranging 0.43–0.59, $P \leq 0.01$ for all) and reduced root distensibility ($r = 0.38$, $P = 0.01$) correlated with AR fraction. Increased PWV in the aortic arch and reduced root distensibility correlated with increased LV mass ($r = 0.38$, $P = 0.01$; $r = 0.35$, $P = 0.02$; respectively).
Conclusions: Aortic root dilatation and minor degrees of AR are shown by MRI in patients with non-stenotic BAVs. In addition, MRI reveals reduced aortic wall elasticity that correlates with the severity of AR and LV mass. MRI may be useful for monitoring of aortic dimensions and aortic elasticity, in conjunction with aortic valve competence and LV function in patients with BAVs.

O2-1
Caval flow reflects Fontan hemodynamics: Quantification by magnetic resonance imaging
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Introduction: Suboptimal Fontan hemodynamics is a multifactorial problem and is dependent on pulmonary artery resistance und ventricular function. We investigated whether flow volume in the inferior (IVC) and superior (SVC) caval veins and flow distribution between SVC and IVC reflect Fontan hemodynamics.
Methods: Flow was measured using phase contrast MRI in 61 patients who underwent different modifications of Fontan operation between 1992 and 2006. Median age at MRI was 12 (3–45) years. Median follow-up was 5.7 (up to max of 14.1 years). Flow volumes in the IVC and SVC as well as combined venous return (cardiac index, CI) and the flow distribution between IVC and SVC were determined and correlated with suboptimal Fontan hemodynamics, particularly with chronic ascites, pleural effusions and patient age.
Results: MRI measurements show a median flow volume in the SVC of 1.1 (0.1–3.4) L/min/m² and in the IVC of 1.8 (0.6–3.2) and total CI of 3 (1.2–5.1) L/min/m². Patients with suboptimal hemodynamics ($n = 8$) had significantly lower flow volumes in the IVC compared to patients with stable hemodynamics (median of 1.5 vs. 1.9 L/min/m², $p = 0.027$). Flow volume distribution in patients with ascites was nonphysiological (dependent on the patient's age), with a significantly lower flow volume in the IVC and increased quotient of the SVC flow volume (0.37 of the combined flow volume in patients with ascites vs. 0.56 in patients with optimal hemodynamics, $p = 0.012$). We found a significant correlation between decreased total CI and higher patient age in the MRI testing ($r = 0.538$, $p < 0.001$).
Conclusions: Fontan hemodynamics can be analyzed noninvasively by MRI. Patients with ascites show reduced flow volume in the IVC and increased flow volume in the SVC in comparison to those with optimal Fontan hemodynamics. Furthermore, the total cardiac index decreases in all patients during the long-term follow-up.

O2-2
Arterialisation of hepatic blood flow in patients with a Fontan circulation
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Introduction: The absence of a sub-pulmonary ventricular pump in the Fontan circulation results in raised central venous pressures, depressed cardiac output, and variable cyanosis, known risk factors in the development of cardiac cirrhosis. Arterialisation of hepatic blood flow secondary to these factors may be a key mechanism

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by which hepatic damage is initiated. We report our observations of arterialised regenerative nodules (indicating arterialisation of hepatic blood flow), and correlate these findings with underlying Fontan haemodynamics.
Methods: Data on 27 Fontan patients (21 atrio-pulmonary, 3 total cavo-pulmonary, 2 right atrial (RA)- right ventricular and 1 Kawashima variant; mean age 25.2 ± 5.7yr) investigated between September 2003–January 2007, were reviewed. Candidate haemodynamic variables (RA pressure, RA saturation, aortic saturation) were examined in relation to the following imaging findings: arterialised nodules, varices, portal venous phase hepatic enhancement characteristics, (normal, reticular and zonal) and portal vein dimensions.
Results: 24 patients having had their primary Fontan at mean 5.5 ± 2.8 yr demonstrated a reticular enhancement with no significant haemodynamic correlation. Mean RA pressure was 14.3 mmHg (± 4.8SD). Mean RA saturation was 67.4% (± 9.7SD) and mean aortic saturation was 93.4% (± 6SD). Patients (7 of 27) with arterialised nodules (size 0.5–3.2cm) had higher mean RA pressures (17.7 ± 5.6SD vs. 13.1 ± 4SD mmHg, P = 0.025), where as their mixed venous saturation and aortic saturation was not significantly different (70%(± 11.9SD) vs. 66.5%(± 9.13SD) and 91.7%(± 9.5SD) vs. 94.1%(± 4.3SD)). Splenic size was abnormal in 9 (mean size 15cm, range 13–20cm). Of 7 patients with extra-hepatic varices, 2 also had lienorenal shunts. Patients with varices had higher mean RA pressures (16.9 ± 6.7SD vs. 13.4 ± 3.8SD mmHg, p = 0.0499) and higher RA saturations (75 ± 5.3SD vs. 65.5% ± 9.7SD, p = 0.016).
Conclusion: The Fontan circulation is almost uniformly associated with an altered liver perfusion as detected by a reticular perfusion pattern in the portal venous phase during CT imaging of the abdomen. In patients with high Fontan pressures, arterialised regenerative nodules commonly arise, marking the onset of arterialisation of liver blood flow, a potent inducer of hepatic fibrosis. Extra-hepatic varices also commonly arise in this setting likely reflecting venous bypass of the higher hepatic resistance.

O2-3
Compliance of the normal-sized aorta in adolescents with Marfan syndrome: Comparison of MR measurements of aortic distensibility and pulse wave velocity
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Purpose: To compare the aortic compliance of the normal-sized aorta of adolescents with Marfan syndrome and healthy controls using MR measurements of the aortic distensibility and the pulse wave velocity.
Materials and Methods: Fourteen patients (median age: 15 [9–21] years) and 11 healthy subjects (23 [12–32] years) were examined on a clinical 1.5 T whole-body MR scanner. The MR protocol included 2D CINE MRI of the aortic distensibility (TrueFISP); and PC-MRI of the pulse wave velocity. All measurements were positioned perpendicular to the descending aorta at the level of the diaphragm for assessing the changes in the aortic cross-sectional areas and additionally above and below this plane for assessing the pulse wave velocity. In addition contrast-enhanced 3D-MR angiography was performed in adolescents with Marfan syndrome to exclude morphologic changes and to prove normal-sized aorta.
Results: Compared with the control subjects, adolescents with Marfan syndrome had significantly decreased distensibility and significantly increased pulse wave velocity (chi^2-test, $p < 0.0002$) using an age-related non-linear regression analysis. The related aortic compliance was significantly decreased (chi^2-test, $p < 0.0002$). There was a good correlation between the two methods ($r = 0.86$). Also a low intraobserver variability was found for both methods (<2%).
Conclusions: MRI allows detecting abnormal elastic aortic wall properties already in the normal-sized aorta of adolescents with Marfan syndrome. Monitoring of these properties could be relevant for evaluating disease progression and treatment options, e.g. indication of elective surgery. For that purpose further studies examining the potential value of compliance measurements for the follow-up and to guide therapy indications are warranted.

O2-4
Strong and Independent Prognostic Value of Peak Circulatory Power in Adults With Congenital Heart Disease
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Introduction: The identification of adult congenital heart disease (ACHD) patients that are at higher risk of death is challenging. Peak circulatory power (CircP, expressed as peak exercise oxygen uptake multiplied for peak mean arterial blood pressure) is a strong predictor of death in adults with acquired heart disease. We sought to establish the distribution and the prognostic value of peak CircP across a wide spectrum of ACHD patients.
Methods: Four-hundred thirty two consecutive ACHD patients of varying diagnosis and 98 healthy controls underwent cardiopulmonary exercise testing at a single laboratory between 1996 and 2005. Patient age was 32 ± 10 years.
Results: A gradual decline in peak CircP was found across the spectrum of congenital heart defects ($p < 0.0001$ at ANOVA). Reduced peak CircP values were associated with presence of heart failure symptoms ($p < 0.0001$), absence of sinus rhythm ($p = 0.010$), and use of antiarrhythmic medications ($p = 0.0013$). At a follow-up of 4.4 ± 2.4 years, 23 patients (5.3%) had died. Peak CircP was a strong predictor of mortality at univariate analysis and the strongest independent predictor of mortality among exercise parameters. A peak CircP? 1406 $mmHg \cdot mlO_2 \cdot min^{-1} \cdot kg^{-2}$ was associated with a 14-fold increase in the risk of death (24% at 5 years of follow-up).
Conclusions: Peak CircP is abnormal across the spectrum of ACHD. Peak CircP appears as the strongest predictor of adverse outcome in ACHD.

O2-5
Predictors of rhythm and conduction disturbances in adult patients with congenital heart diseases.
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Introduction: Abnormal heart structure and performed cardiac surgery may results in rhythm and conduction disturbances (RCD) in adults with congenital hart disease (CHD).
Aim of the study: The assessment of risk factors for RCD in adults with CHD during long-term outcome.
Material and method: 1304 patients (P) (586 men) aged 18–72 (mean 29.4 ± 10), were studied and observated during 1–10 years (mean 3.52 ± 1.83) (1995–2004). In P 25 different types of CHD were diagnosed – 13 simple: ASD, VSD, PDA, bicuspid aortic valve, pulmonary stenosis, subvalvular aortic stenosis, VAC, partial anomalous pulmonary venous connection, Marfan's syndrome, mitral insufficiency, idiopathic dilatation of the pulmonary trunk, additional vena cava superior sinistri, Wiliamas syndrome and 12 complex CHD: coarctation of the aorta, Fallot's tetralogy, ASDI, CAVC, Ebstein syndrome, univentricular heart, CCTGA, DTGA, DORV, pulmonary atresia, coronary fistula, Bland White Garland syndrom. Analyzed RCD: all atrio-ventricular conduction blocks, nodal rhythm, sinus arrest ≥2 sek., advanced bradycardia (< 30 bpm) during sleep and <40 bpm during day.
Anatomical complexity of the CHD, performed cardiac operation, heart failure degree (NYHA > I), cyanosis presence, age and gender were assessed during first visit. Kaplan-Meier curve were estimated, log-rank tests to compare curves, and Cox proportional hazards model for assessing multivariate associations between RCD occurrence and risk factors were calculated.
Results: RCD were observed in 11.1% of P. The probability of their occurrence after 2 years – 4.0%, 5 years – 10.0%. Univariate analysis: probability of RCD occurrence is higher in men (p = 0.006), P with complex CHD (p = 0.00001), NYHA > I class (p = 0.0002), cyanosis (p = 0.002). The risk factors for RCD in all patients: anatomical complexity (HR = 3.1), male gender (HR = 1.58). Risk factors in complex CHD: male gender (HR = 2.9), heart failure (HR = 2.2). Risk factor in simple CHD: cyanosis (HR = 5.9). Previous cardiac operation and age did not have the influence on RCD occurrence
Conclusions: Anatomical complexity, male gender have essential prognostic significance for RCD.
The risk factors for RCD occurrence are different in groups with simple and complex CHD.
Male gender and heart failure is a significant prognostic factor in complex CHD. In patients with simple CHD the risk increases with the cyanosis presence.

O2-6
Health Related Quality of Life of Adults with Congenital Heart Defects
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Introduction: Most of the children with congenital heart defects (CHD) nowadays survive into adulthood. However, as a prolongation of life does not necessarily mean a good quality of life, the measurement of the quality of life of adults with congenital heart disease gains an essential value for the long-term surveillance.
Methods: A questionnaire was sent to 1,091 adult patients enrolled in the National Register for Congenital Heart Defects in Germany. The health related quality of life was measured using the SF-36. After correction for age and sex the results are compared with the data from the German National Health Survey (n = 7.066).
To examine the importance of the degree of severity of the diagnosis we compared the results of a group of adults with ASD (n = 176), TGA (n = 189), TOF (n = 181) with a group with highly complex lesions (n = 201; PA, TrA, TAC, DIV, PHT).
Results: Compared with the healthy population we found major differences in the dimensions 'physical limitations' (77 vs. 86), 'physical role functioning' (75 vs. 83), 'body pain' (84 vs. 68) and 'emotional role functioning' (80 vs. 90). Moderate differences were detected in the dimensions 'social functioning' (84 vs. 87), 'vitality' (58 vs. 60) and 'general health perception' (61 vs. 67). No statistically significant difference could be found in the dimension 'psychological wellbeing' (73 vs. 73).
In the comparison of the different diagnostic groups, we found a significant difference with respect to the physical dimensions, but not with respect to the psychological wellbeing. The presence of cyanosis has a remarkable influence on all scales but the psychological wellbeing.
Conclusion: In this big sample health related quality of life of adults with CHD differs from the healthy population in all scales but the psychological subscale. The biggest difference can be found with respect to the physical domains and the emotional role functioning. Patients perform well on the scale for psychological wellbeing. Cardiac diagnosis, patient's sex and age seem to affect QoL. More complex diagnoses are exposed to a relatively higher burden, but the highest risk factor for a reduced quality of life is with no doubt the presence of cyanosis.

O3-1
Effect of Beta-Blocker Therapy in Children with Dilated Cardiomyopathy: A Single Centre Retrospective Study
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Background: Beta-adrenergic blockade is a standard therapy in adult patients with LV dysfunction.
Few data exists about the efficacy of ß-blockers in children with dilated cardiomyopathy.
Aim: This retrospective study sought evidence for the efficacy of ß-blocker therapy in children with DCM using transplant free survival (TFS) and EF as end points.
Methods: We reviewed the medical records of all 29 DCM patients referred to our hospital from 2000 to 2006 with the following characteristics: a) endomyocardial biopsy negative for myocarditis, b) EF <40% at presentation, c) standard anticongestive therapy. Twelve of these 29 children (group A) were treated with anticongestive therapy plus a ß-blocker, while 17 (group B) received anticongestive therapy alone. ß-blocker therapy was started a median of 11.7 months (range 0–50) after the diagnosis. Statistical analysis was performed with a two-sample t test. Survival curves were compared with the log-rank test.
Results: All the children, except one treated with metoprolol, received Carvedilol, starting at a dosage of 0.1 mg/Kg/day and increasing to a mean of 0.7 ± 0.2 mg/Kg/day. The two groups did not differ in age, weight and ejection fraction at diagnosis (p > 0.05). Over the FU the EF increased similarly in the two groups (from a median of 27 to 47% (p 0.0008) in group A and from a median of 29.5 to 38% (p 0.004) in group B. Four children in the group A (33%) reached an end-point (59%) compared with 10 (59%) of group B. The survival curves differed significantly (p 0.02).
Conclusions: In our children affected by DCM the addition of a ß-blocker to anticongestive therapy did not influence the change in EF during follow-up. Although the TFS curves differed significantly for the 2 groups, most percentage of the endpoints seen in group B occurred during the first year. Because ß-blocker therapy was started late after diagnosis (median 11.7 months), it is possible that the treated group is biased for patients like to survive. Although our data do not convincingly show a benefit of

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ß-blocker therapy in our children with DCM, we can not exclude the possibility.

O3-2
Utility of Signal-Averaged Electrocardiography in Pediatric ARVC

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Introduction: Signal-averaged electrocardiography (SAECG) is reported to have a moderate sensitivity (SENS) and high specificity (SPEC) for ARVC in adults. We sought to identify SENS and SPEC of SAECG in children.
Methods: Patients presenting with LBBB VT or a family history of ARVC underwent noninvasive assessment for Task Force (TF) criteria for ARVC, including SAECG. Those identified with an additional ARVC criterion underwent invasive assessment. SAECGs were filtered at 40–250Hz. Z values for filtered QRS duration (QRSDf), high frequency low amplitude signal duration (HFLA) and root-mean-square voltage of the terminal 40 msec (RMS40) were calculated based on published regression equations from a group of 139 normal children aged 5–15 years, who were also used to assess SPEC of individual and combined SAECG criteria. Where ARVC patients had sequential SAECGs during follow-up, changes in Z value were calculated on a per year interval (dZ). Outcomes of death or appropriate defibrillator shock (DEATH/SHOCK) were identified.
Results: SAECGs were available in 36 children aged 4.8 to 18.0 (13.9 ± 3.2) years who met TF criteria for ARVC. Average noise was 0.26 ± 0.09 mV. SAECG parameters were > 2 S.D. for QRSDf in 9/36 (25%), > 2 S.D. for HFLA in 11/36 (31 < −2 S.D. for RMS40 in 9/36 (25%). Any abnormal SAECG parameter was present in 17/36 (47%) children, including 6 who had normal unfiltered QRS duration (QRSDu) on standard ECG. Only 11/139 (8%) control children had any abnormal SAECG parameter. DEATH/SHOCK occurred in 4/36 patients, and was predicted by a more prolonged HFLA (Z value = 3.99 ± 4.36 vs. 1.65 ± 2.11, $p < 0.02$) and lower RMS40 (Z value = $- 2.33 \pm 1.85$ vs. -1.16 ± 0.98, $p < 0.01$) Patients with DEATH/SHOCK tended to have a more rapid negative change (dZ) in RMS40 (-0.58 ± 1.23 vs. -0.03 ± 0.21, $p = 0.1$).
Conclusions: In these children, SAECG has a 47% SENS and 92% SPEC for ARVC, and identified additional children with ARVC whose QRSDu on standard ECG is normal. SAECG is feasible in children being assessed for ARVC, with SENS and SPEC approaching that seen in adults assessed for ARVC. In this limited sample size, SAECG parameters and their trends appear to predict outcome.

O3-3
Quality of life after Fontan and TCPC-operation

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Background: The patients born with a univentricular heart are nowadays palliated with surgery leading to Fontan circulation. The overall mortality of the operated patients has improved during years. Recently an important question has been raised, to what extent health-related quality of life (HRQoL) is gained with the operation.
Material: The study included all 83 patients over eight years of age having Fontan circulation, and living in Finland in November 2005.
Methods: The HRQoL of patients was measured by the 15D instrument and it's versions for adolescents (16D) and children (17D). The questionnaires were mailed to the patients. The results were compared to age- and gender-standardised general population. Statistical significance of the difference was assessed with the t-test.
Results: Of the 26 adults 19 (73%) returned the questionnaire. The mean age of the respondents was 22.5 (range 16.3–29.5) years, and nine of them were men. The patients' overall mean HRQoL score was lower than that of controls (n = 947), 0.92 and 0.97, respectively ($p < 0.001$). Patients reported problems especially in moving, breathing, usual activities and sexual activity.
Among the adolescents the response rate was 89% (24/27 pts). The mean age of the respondents was 13.8 (range 12.1–15.9) years, and half of them (12) were male. The overall mean HRQoL score was equal to that of controls (n = 239), 0.93 vs 0.95, respectively ($p = 0.081$). However, patients had difficulties especially in breathing, eating, school and making friends.
Of the 30 children 26 (87%) returned the questionnaire, and 13 of them were male. The mean age was 10.1 (range 8.3–11.9) years. The overall HRQoL score was 0.88 among patients, and 0.94 in controls (n = 244), $p < 0.001$. Patients experienced difficulties especially in breathing, eating, speech, elimination, school, making friends, and concentration.
Conclusion: The overall HRQoL of patients born with a univentricular heart was quite good, although slightly worse than that of controls. The patients experienced some difficulties in their every day life. Most problems occurred on dimensions where they were expected, for example in breathing. The surprisingly good subjective Qol in these patients stresses the importance of patient-derived measures in assessing treatment outcome.

O3-4
Low incidence of inappropriate or ineffective ICD therapy in children and adolescents with implantable cardioverter defibrillators. Results of a single center medium term study

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Introduction: Reported incidence of inappropriate or/and ineffective therapies are matter of concern probably limiting lifesaving therapies in the young persons needing implantable cardioverter defibrillators (ICD).
Methods: The data of all of our patients (pts) with previously implanted ICD were retrospectively analyzed regarding shock therapies and major complications.
Results: 27 pts underwent ICD implantation between January 2001 and December 2006. The underlying cardiac disorders included congenital heart disease (37%), long QT-syndrome (18.5%), Brugada-syndrome (7%), hypertrophic cardiomyopathy (26%) and others (11%). Implant indications were fast VT (14.8%), torsade or documented VF (18.5%), syncope (26%), hemodynamic unstable VT (22%), resuscitation (11%) and others (7%). The mean age at implantation was 17 years (range 8–36). During operation there were no major complications. All pts had transvenous screw in electrodes implanted in the RV except one pt with LV implantation S/P Senning. Dual chamber ICDs were implanted

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in 16, biventricular ICDs in 2 and single chamber ICDs in 9 pts. 51 shocks (range 1–34) were delivered in 9 pts during a cumulative follow up of 881 months (range 1–172). All therapies were delivered for tachycardias, 49 shocks (96%) appropriate for VT/VF. In all but one pts the very first shock terminated the arrhythmia, shock delivery in a pt with end-stage HCMP was ineffective but VF terminated spontaneously afterwards. One pt with LQTS with a single chamber ICD and one pt with Brugada-syndrome with a dual chamber ICD received inappropriate DC-shocks, each of which caused by atrial flutter.
Conclusions: 1. ICD-implantation is a highly effective therapy for lifethreatening ventricular dysrhythmias in children and adolescents with or without congenital heart disease. 2. Using standard guideline ICD indication criteria our data do not show the previously elsewhere reported high incidence of inappropriate shock delivery. 3. We did not find evidence for lead problems using Medtronic Sprint or Guidant Endotak screw in leads. 4. In atrial flutter dual chamber system algorithms did not prevent inappropriate ICD therapy, so SVT distinction algorithms should be further improved.

O3-5
Estimation of fetal atrioventricular (AV) time intervals by pulse Doppler and Doppler tissue echocardiography
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Background: Data from literature shows that Doppler tissue imaging allows more accurate measurement of atrioventricular (AV) time intervals than pulse Doppler in fetus.
Objective: To assess gestational age-specific values of normal fetal AV time interval by pulse-wave Doppler (PD) methods and Doppler tissue imaging (DTI).
Methods: Examinations were performed using Sequoia 512 in 64 healthy singleton fetuses at 17 to 34 weeks of gestation. PD-derived AV intervals (PD-AV) were measured from left ventricular inflow/outflow view using convex 3.5–6 MHz probe. DTI-derived AV intervals (DTI-AV) were measured from atrial contraction (Aa) to isovolumic contraction (IV) at the base of right ventricular free wall using pediatric cardiology probe of 2.5–7 MHz.
Results: In gestational age group of 17–20; 21–24; 25–29 and 30–34 weeks mean values of PD-AV were: 110.6; 114.5; 118.9; 126.4 ms ($p = 0.001$) and of DTI-AV: 93.9; 96.2; 99.6; 102.8 ms respectively ($p = 0.026$). PD-AV and DTI-AV were negatively correlated with heart rhythm ($p = 0.01$).
Conclusions: DTI-derived AV intervals were shorter than PD-derived AV intervals. AV duration measured by PD and DTI was positively correlated with gestational age and negatively correlated with heart rhythm. This indicated that mechanical intervals are not the same as electrical intervals, what is necessary to keep in mind in suspicion of prolonged interval in fetuses, for instance prolonged QT. Further studies are needed to make those methods more specific.

O3-6
The Amplatzer® Membranous VSD Occluder and the Vulnerability of the Atrioventricular Conduction System
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Transcatheter ventricular septal defect closure with the Amplatzer® Membranous VSD Occluder has yielded promising initial results but conduction disturbances including complete heart block have been reported. We report our experience with the Amplatzer® occluder on 35 patients with median age 4.5 years, angiography defect size 4.4 ± 1.1 (3.0–8.0) millimetres and occluder size 4–12 millimetres. Over median follow-up of 2.5 years, complete closure rate was 87% and 91% at 1 and 2 years respectively while 2 patients required post-procedure surgical defect closure. Persistent procedure-related regurgitation was observed in 3 patients in the tricuspid and 6 patients in the aortic valve. Procedure-related conduction abnormalities were noted in 7 patients (20%), transiently in 1 and permanently in 6 patients (17%), in one of which it progressed after 6 months from left bundle branch block to intermittent Mobitz II second-degree atrioventricular block associated with expansion of the occluder. Transcatheter closure of perimembranous ventricular septal defects with the Amplatzer® occluder proved effective with limited complications in this study but the immediate and progressive conduction disturbances observed along with the proximity of conduction tissues to the defect rim stress the importance of larger and longer studies to assess safety of this procedure.

O4-1
Autopsy protocol for prenatal cardiovascular anomalies – a substrate for successful cooperation between pathology, anatomy and prenatal cardiology specialists
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Introduction: The role of prenatal autopsy in assessment of cardiac malformations diagnosed prenatally seems to be well established. Detailed post-mortem examination requires well-trained specialists and time, so it can be impossible to do it during routine autopsy of non-fixed fetal cadaver. Pathologists who are not trained in cardiovascular anatomy and pathology are often focusing on standard dissection methods. Frequently, autopsy reports do not include segmental description of the heart and important facts regarding anatomy of cardiovascular system and other associated anomalies. In our institution we developed a protocol for examination of these cases, including procedure of removal of thoracic and abdominal viscera. It assures preservation of all anatomical structures necessary in cases of complex cardiovascular anomalies.
Methods: We performed autopsies 20 fetuses using our algorrhitm. After removal, organ blocks were fixed in 4% formaldehyde solution and dissected carefully under magnification of dissection microscope when needed. Segmental analysis of cardiac anatomy and detailed description of cardiac structures, as well as information about anatomy of relevant extracardiac structures were mentioned in autopsy report. Prior to autopsy prenatal cardiologist and cardiac anatomist reviewed all echocardiographic examinations in order to plan the most appropriate method of opening cardiac chambers and dissection of specimen. After the report has been prepared, echocardiographic examinations were reviewed again and compared with autopsy findings.
Results and conclusions: Presented form of cooperation between prenatal cardiologist, anatomist and pathologist confirmed its

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usefulness especially in cases of visceral heterotaxy syndromes, where it has helped to reveal details of anatomy, which has not been visible on echocardiography and standard autopsy. We believe that such procedure assures the highest relevance of post-mortem examination and helps to understand anatomy of complex anomalies. In order to illustrate the value of our protocol, we present some cases of complex cardiovascular anomalies from our material.

O4-2

4D Fetal Echocardiography in Prenatal Diagnosis of Congenital Heart Disease

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Objective: The development of new echocardiographic techniques has been a corner stone of prenatal diagnosis of congenital heart disease (CHD); real-time 3D colour Doppler echocardiography (4D) is no exemption. We used a systemic real time 4D (STIC) to image the fetal heart. The aim of the study was to prove that 3D cardiac volumes obtained from the 4-chamber view can adequately visualise the different cardiac parts and correlated this with 2D Doppler studies of CHD to examine its function as a prenatal screening tool for CHD.
Methods: This study included foetal STIC investigations performed between 2004–2005. Acquisition varied in angle between 20–30° and took 10–15 seconds. Two groups were analysed, a normal fetal population and a group of fetuses with CHD. The first was analysed to evaluate the image availability of the different cardiac structures, in the second the efficacy of diagnosis based on 4D evaluations was compared to 2D results and post-natal outcome.
Results: 47 cardiac volumes with the 4-chamber view as starting point during the acquisition were analysed in fetuses with normal cardiac anatomy (30 between 17–24, 17 between 25–36 weeks GA). An inexperienced investigator with a basic formation in echography reviewed the data for the views required for a standard fetal cardiac evaluation. Visualisation in 17–24 and 25–36 weeks GA: 4-Chamb 96–100%, Septum 96–100%, AV-valves 100%, Pulm. Art 91–100%, Ao 91–100%, Duct. Art. 57–79%, Ao arch 35–21%, Pulm Ven. 25–21%.
The 10 STIC studies of fetuses with CHD obtained between 14–36 (mean: 27) weeks GA: Cardiac analysis was possible in 95%, complete diagnosis in 74%, partial in 16%, requiring modification in 5% and unobtainable in 5%. In 1 case an associated diagnosis of complex CHD, which was missed before, could be diagnosed and was confirmed postnatally.
Conclusion: The use of STIC volumes allows inexperienced investigators to obtain the different views necessary for a complete cardiac exam in an offline fashion. Secondly the achieved good correlation between direct 2D and indirect offline STIC diagnosis suggests that this new technique can be reliably used for the prenatal screening of CHD in the hands of experienced fetal cardiologists.

O4-3

Ingestion of Herbal Teas and Grape Juice During Pregnancy is Associated to Fetal Ductal Constriction: a Clinical Approach

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Background: Inhibition of circulating prostaglandins may promote fetal ductal constriction, especially in late gestation. The hypothesis that maternal consumption of widely used polyphenol-rich substances, such as herbal teas and grape juice during pregnancy could be associated to fetal ductal constriction, as a result of prostaglandin inhibition-mediated antiinflammatory effects of the catechin components of these beverages, has been formulated. This study shows clinical evidences pointing toward corroboration of this hypothesis.
Methods: A review of a series of 92 fetuses with ductal constriction has been performed. A cohort of the last 34 cases, sequentially seen after this hypothesis was brougt up, in late 2005, made up the basis for this report. Differences were analyzed by Wilcoxon test and associations were tested by Fisher's exact test.
Results: In 24 fetuses beyond 30 weeks out of 34 with ductal constriction (77%), there was evidence of maternal ingestion of polyphenol-rich beverages (green tea, mate tea, Indian tea, boldine tea and grape juice). Immediate discontinuation of these substances was reccomended, and a control echocardiogram was obtained after 1 to 3 weeks in 11 fetuses. Of these, 10 (91%) showed important improvement or complete recovery of the signs of ductal constriction. A decrease in mean ductal systolic velocity (1.74 ± 0.25 m/s to 1.34 ± 0.29 m/s, $p = 0.005$) and mean diastolic velocity (0.38 ± 0.09 m/s to 0.22 ± 0.03 m/s, $p = 0.0001$, with an increase in mean ductal pulsatility index (1.60 ± 0.43 to 2.14 ± 0.28, $p = 0.001$) were observed. Only one of the 7 fetuses with other triggering factors (nonsteroidal antiinflammatory drugs, corticosteroids, paracetamol) improved in the same period. Out of the 11 fetuses with improvement in the control echo, 10 (91%) were in the polyphenol group. There was significant association between improvement and polyphenol discontinuation ($p = 0.005$).
Conclusion: Maternal ingestion of substances containing polyphenols is very common in fetuses with ductal constriction and discontinuing these beverages results in improvement in the vast majority of cases.

O4-4

Longitudinal Doppler study of the embryonic heart in low risk pregnacies and their follow up

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Objective: The aim of this study was to describe the normal fetal cardiac and hemodynamic development in normal 4th–10th week gestation pregnancies for better understanding of normal cardiac hemodynamics during the period of embryonic heart development. Further, we tried to find any abnormal Doppler signals or markers predicting abnormal heart development or miscarried pregnancies.
Materials and methods: One hundred women with singleton, uncomplicated pregnancies were prospectively studied with transvaginal ultrasound, pulsed and color Doppler. Heart size, heart rate, inflow and outflow waveforms with valve signals were documented. The proportion of the cardiac cycle of isovolumetric relaxation (IRT%) and contraction time (ICT%) as well as Tei index were calculated. Peripheral flows at ductus venosus, umbilical artery and vein were also analyzed.
Results: Eighty nine % of studies were successful. Heart size and the fetal heart rate showed a positive correlation with increasing gestational age $R = 0.80$ ($p < 0.000001$), $R = 0.76$ ($p < 0.000001$),

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respectively. Mean heart size at 6 weeks was 1.13+0.3 mm and mean fetal heart rate was 113+20 beats/min compared to 3.95+0.6 mm and 171+6 beats/min at 10 weeks. Inflow waveform was monophasic (atrial contraction) in all cases from 6 to 9 weeks. Fourteen pregnancies (10%) miscarried between 6–12 weeks of gestation and the heart exams were characterized by increased IRT% compared with the survivors. Six (43%) of them had abnormal heart rhythm. In survivors, IRT% decreased from 7 to 8 weeks – from 31.1 + 10.2% to 20.2 + 5.7% ($p < 0.0001$). ICT% decreased from 18.0 + 4% of the cardiac cycle at 8 weeks to 12.2+6.3% at 9 weeks ($p < 0.0008$) – (after heart development period). Aortic stenosis was recognized in 1 case, confirmed in neonatal period.
Conclusions: Doppler examination of the fetal cardiac function is possible after 5 weeks of gestation. After 8 weeks of gestation, the fetal heart is morphologically mature but has not yet achieved effective myocardial compliance. The embryonic human heart is dependent on the atrial contraction for ventricular filling. Non-survivors manifest myocardial dysfunction. Bradycardia or tachycardia was a poor prognostic factor.

O4-5
Comparison of first line transplacental antiarrhythmic therapies in fetal tachycardia
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Background: Fetal tachycardias can lead to severe hemodynamic compromise and even death. Transplacental therapy by maternal antiarrhythmic drug administration has significantly improved clinical outcome. The objective of this study was to compare the efficacy and safety of flecainide, sotalol and digoxin as first line maternal drug therapies for fetal supraventricular tachycardia (SVT) and atrial flutter (AF).
Methods: From 1998 to 2006 95 consecutive cases of fetal tachycardia from two centers were retrospectively evaluated.
Results: SVT was diagnosed in 78% (n = 74) and AF in 22% (n = 21). The SVT group included 95% atrioventricular re-entrant tachycardia (AVRT) and 5% ectopic atrial tachycardia. 24 pts (25%) had hydrops, all in the SVT/AVRT group (34%). Cardiac anomalies were present in 7%. Transplacental therapy as first line therapy was given in 65 pts (68%), flecainide+/−digoxin (n = 15), sotalol+/−digoxin (n = 31) or digoxin only (n = 19). In the presence of hydrops digoxin was only given in 1 pt (4%). Median onset of tachycardia was 29+5 wks in the treatment group and 35+3 wks in the untreated group. (Partial) success rate of first line therapy within 5 days was 41% in the hydropic group and 57% in the non-hydropic group. (Partial) success rate did not significantly differ between the first line drug groups. However, conversion to sinus rhythm occurred earlier in the flecainide group (all <3 days) than in the sotalol or digoxin group. After first line drug failure subsequent treatment strategies included alternative transplacental therapy (flecainide/sotalol/amiodarone), direct fetal therapy (amiodarone) or delivery. Overall mortality was 9%, tachycardia-related deaths occurred in 4 pts (4%). Other causes of deaths (5) were related to tumors, metabolic disease or perinatal complications. Post-hemorrhagic hydrocephalus (no neurological sequelae) occurred in 2%. None of the fetuses had documented side-effects of the drugs given.
Conclusions: Efficacy and safety of transplacental drug therapy with flecainide and sotalol are comparable in both hydropic and non-hydropic fetuses with SVT or AF. In the absence of hydrops digoxin monotherapy appeared to be just as effective. Alternative treatment strategies should be planned timely after first line therapy failure.

O4-6
3 Vessel View Z-score calculator: a practical tool for increasing detection of fetal coarctation of the aorta at the first screening examination
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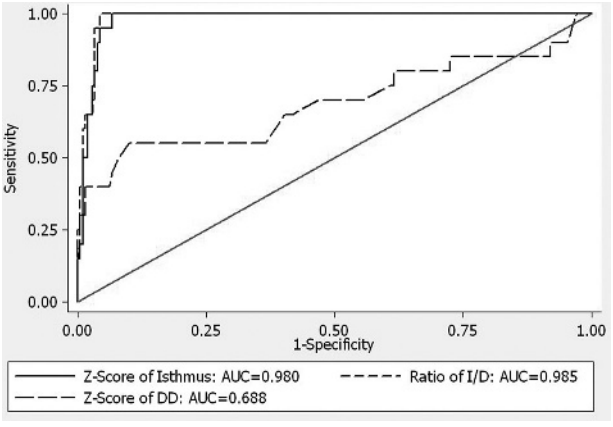
Introduction: Prenatal diagnosis of isolated coarctation of the aorta (CoA) suffers from high false positive and false negative rates. Suspicion is raised by disproportion at four chamber and/or great arterial views but this is not quantitative and may be less dramatic in the presence of associated ventricular septal defect. Missed cases of CoA result in significant morbidity & mortality so improvement in detection is important.
Methods: We tested the ability of Z-scores of aortic isthmus (AoI) and ductal diameter (DD) and the AoI:DD ratio, measured in the three vessel and tracheal view (3VV), to separate fetuses with suspected CoA from normals at the first screening examination. AoI is measured immediately proximal to the insertion of the arterial duct and DD immediately before it enters the descending aorta in the 3VV.
We studied 31 fetuses with suspected CoA referred at 23+5 (range: 17+3 to 34+4) weeks' gestation and archived data from 200 normal consecutive singleton fetuses referred for cardiac scan at median 22+0 (range 15+4 to 38+4) weeks' gestation. Receiver Operator Characteristic (ROC) curves were created from the first examination.
For our previously derived Z-score calculator, see: Pasquini L. et al. "Z Scores of the fetal aortic isthmus and duct: an aid to assessing arch hypoplasia". UOG 2007, in press.
Results: There was complete separation of the Z-scores of the normals and cases requiring surgery (16/31) or postnatal surveillance for arch hypoplasia (4/31). ROC curves of AoI and AoI:DD had an excellent Area Under Curve of 0.98 and 0.99 respectively, whereas DD alone was less powerful at 0.69.
Eleven cases (11/31) were normal postnatally (i.e. false positive at first screening), however serial isthmal Z-scores improved during pregnancy in most to >−2 while those requiring surveillance or surgery remained <−2 with positive Z-scores for the duct.



Conclusions: The 3 Vessel View Z-score calculator is a relatively simple and practical tool for identifying suspected fetal coarctation of the aorta. Furthermore, serial Z-scores help to reduce any increase in false positives.

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O5-1

Outcomes in children with pulmonary arterial hypertension: Dutch experience

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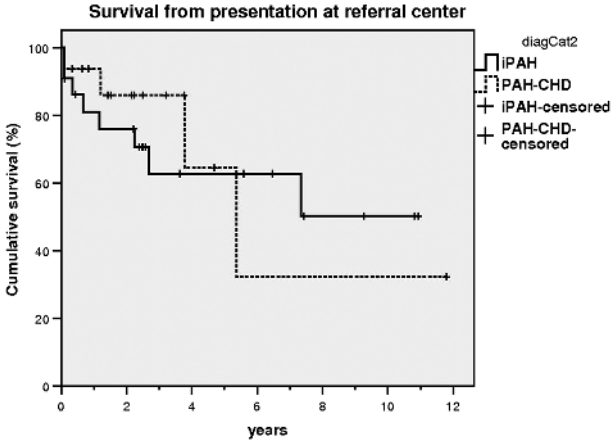
Introduction: Data on treatment of children with pulmonary arterial hypertension (PAH) are limited compared to adults with PAH. We investigated the outcome of children with different types of PAH treated with various PAH medications.

Methods: Between 1994 and 2006 a cohort of 47 children was referred to the Dutch referral center for diagnosis and treatment of paediatric PAH. Characteristics of PAH at presentation and its treatment were assessed, including transcutaneous oxygen saturation (TcSO2), World Health Organization functional class (WHO class) and hemodynamics. Subsequently, survival was analyzed.

Results: Mean age at presentation was 6.9 ± 5.7 years (range 0.1–17.4). Nine (19%) children had a transient form of pulmonary hypertension (PH) and 38 (81%) chronic progressive PAH (PAH). Of the latter patients, 22 (58%) had idiopathic PAH (iPAH) and 16 (42%) PAH associated with congenital heart disease (PAH-CHD). At baseline, patients in the total PAH group were in WHO class I/II/III/IV (n = 2/7/18/11), mean TcSO2 was 91 ± 8%, mean pulmonary arterial pressure was 56 ± 20 mmHg. Baseline WHO class and hemodynamics were similar in iPAH and PAH-CHD patient groups. TcSO2 was lower in the PAH-CHD group (p = 0.007). Median follow-up was 2.4 years (range 0.1–11.8). Treatment consisted of calcium channel blockers (n = 10 iPAH), flolan (n = 8 iPAH, n = 1 PAH-CHD), bosentan (n = 14 iPAH, n = 13 PAH-CHD) and sildenafil (n = 2 iPAH, n = 1 PAH-CHD). One, 3, 5 and 7 year survival rates were respectively 83%, 71%, 65% and 48% (PAH), 76%, 63%, 63% and 50% (iPAH) and 86%, 86%, 64% and 32% (PAH-CHD) (Figure). Decreased survival was associated with worse WHO class at presentation in the total PAH group (p = 0.02) and iPAH group (p = 0.002), but not in the PAH-CHD group. Higher mean right atrial pressure (p = 0.059) and lower systemic venous saturation (p = 0.006) were associated with decreased survival in the iPAH group, but not in the PAH-CHD group.



Conclusions: Survival of paediatric iPAH patients appeared higher compared to reported historical data before the introduction of new PAH drugs. Baseline characteristics as well as survival were similar for iPAH and PAH-CHD patients. However, baseline predictors for survival in our iPAH patients were not predictive in our PAH-CHD patients.

O5-2

Long-term effect of bosentan in patients with pulmonary arterial hypertension associated with systemic-to-pulmonary shunt: does the beneficial effect persist?

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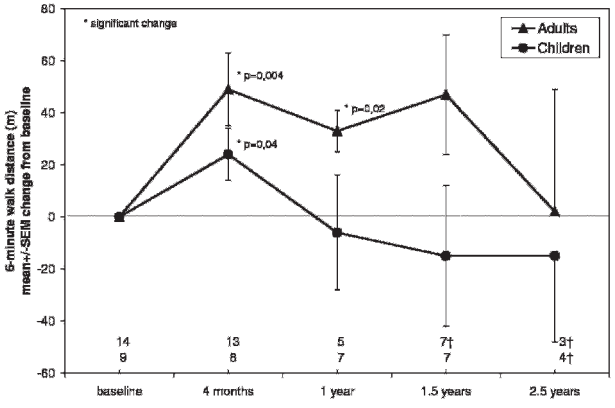
Introduction: In contrast to short-term data, data on long-term response to bosentan (dual endothelin receptor antagonist) in patients with pulmonary arterial hypertension (PAH) in congenital heart disease (CHD) associated with systemic-to-pulmonary shunt are scarce.

Methods: We studied bosentan efficacy in 23 patients (14 adults, 9 children) with the disease at short (4 months), and long-term follow-up (through 2.5 years). World Health Organization functional class (WHO class), transcutaneous oxygen saturation (TcSO2) and 6-minute walk distance (6MWD) were assessed at baseline, 4 months, 1 year, 1.5 years and at latest follow-up (median 2.5 years).

Results: Male:female ratio was 1:3. Mean age at bosentan initiation was 29.8 ± 17.6 years (range 4.7–59.2). Diagnoses included ASD (n = 6), VSD (n = 4), PDA (n = 2), VSD ± ASD ± PDA (n = 7), other complex heart defects (n = 4). Four patients had a closed shunt. At baseline, the total cohort was in WHO class II/III/IV (n = 3/14/6), mean TcSO2 was 88 ± 8%, mean 6MWD was 380 ± 111 m. Median follow-up was 1.8 years (range 0.04–3.4). At 4 months follow-up, WHO class and 6MWD significantly improved (WHO class II/III/IV n = 8/12/2 p = 0.005; mean 6MWD increase 39 m, 95% CI 20–59 m p = 0.0001). During long-term follow-up, WHO class improvement persisted in the total cohort, whereas 6MWD declined. This decline was most pronounced in the children, whereas in the adults 6MWD improvement lasted longer (Figure).



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No change from baseline was seen in TcSO2. Three (13%) patients died, 1 (4%) discontinued bosentan and 5 (22%) required additional sildenafil therapy (of whom 1 eventually died). One and 2-year freedom from loss of beneficial bosentan effect (death, lung or heart lung transplant, atrial septostomy, discontinuation of treatment, requirement of additional PAH therapy: epoprostenol, treprostinil, sildenafil, or decline in 6MWD) was respectively 68% and 36% for the total group, 77% and 47% for the adults, and 56% and 22% for the children.
Conclusions: Our experience with bosentan in patients with PAH associated with systemic-to-pulmonary shunt suggests short-term improvement in WHO class and 6MWD, followed by stability in WHO class and decline in 6MWD at long-term follow-up. Freedom from loss of beneficial bosentan effect decreased progressively over time. The decline was most pronounced in the paediatric patients.

O5-3

Decrease of interventricular Dyssynchrony after cardiac resynchronization therapy (CRT) in patients with congenital heart disease (CHD) is associated with improvement of ventricular function and physical performance

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Background: Due to the limitations of conventional methods and the heterogeneous morphologies identification of patients with heart failure and CHD whom might benefit from CRT is difficult. TDI derived Strain allows quantitative assessment of regional electromechanical coupling.
Patients and methods: 29 patients with DCM (n = 7) and CHD (n = 22) (TOF 5, ISTA 3, Ebstein 2, AS 1, VSD 1, DORV 1, AI 1, cc-TGA 8) who identified to have systemic ventricular dyssynchrony and underwent subsequently cardiac resynchronization therapy (CRT) were examined before, immediately after and every three months during a follow-up period of one year, by conventional and TDI Echocardiography (Vingmed, Vivid 7). In an apical four chamber view using high frame rates (180–250bps) regional deformation (Strain %) was analyzed. The time interval from the onset of QRS-Complex to the maximum Strain, as marked by the aortic valve closure was assessed for three segments of the left and the right lateral free wall, as well as the interventricular septum.
Results: Biventricular pacing lead to a significant reduction of interventricular delay between LV and RV ($p < 0.05$), immediately after CRT as well as during the follow-up period. The decrease in time to peak of myocardial deformation was associated with increase in EF of the systemic chamber, increased diastolic wall velocities (A-Wave) and improvement of exercise capacity (maxVO2).
Conclusion: Interventricular Dyssynchrony can be quantified by TDI derived Strain and seems to be a useful tool for Identification of Patients who might benefit from CRT, as well as for assessment of their response to therapy.

O5-4

Outcome factors of idiopathic dilated cardiomyopathy in children. A follow-up review over twenty years

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Background: Idiopathic dilated cardiomyopathy in children has a high mortality rate by heart failure and final treatment is cardiac transplant in great percentage of children. Nevertheless, there is not a score, which could predict death and that could help in cardiac transplant indication.
Objective: To review the follow-up in the last 22 years of 142 children with idiopathic dilated cardiomyopathy and from that to propose a new prognostic score that predicts risk of death in this group of children.
Patients and Method: A twenty-two years review of 142 consecutive children with idiopathic dilated cardiomyopathy (36 deaths) was undergone. The criteria for inclusion were congestive heart failure, cardiomegaly, and left ventricle systolic dysfunction. Based on Cox's analysis, a new prognostic score index using clinical and laboratorial data was proposed.
Results: The mortality predictors were function class IV at presentation ($p = 0.0001$), dyspnea ($p = 0.0096$) and small pediosous pulses ($p = 0.0413$). In chest x-ray, they were maximum cardiothoracic ratio ($p = 0.0001$) and pulmonary congestion ($p = 0.0072$). In electrocardiogram, right atrium overload ($p = 0.0118$), ventricular arrhythmias ($p = 0.0148$) and heart rate ($p = 0.0275$). In echocardiogram, mitral regurgitation grade 3–4 ($p = 0.0017$), left atrium/aorta ratio ($p = 0.0001$) and left ventricle ejection fraction ($p = 0.0266$). Multivariate analysis showed that the independent predictors factors were maximum cardiothoracic ratio ($p = 0.0001$), left ventricle ejection fraction ($p = 0.0013$), mitral regurgitation grade 3–4 ($p = 0.0017$), function class IV at presentation ($p = 0.0028$) and ventricular arrhythmias ($p = 0.0253$). A score was built with eight maximum points. One child with a score five or higher has a relative risk of 4.00 (confident interval of 95% 2.82 to 5.69 – chi-square = 52.68) against other with a score 4 or less ($p = 0.0001$). This score had a sensitivity of 94.4%, specificity of 76.4% and fitted ROC area 0.881 ± 0.028.
Conclusion: Children, who have five or higher points in this new score, should be considered for early heart transplantation, if no improvement was observed in clinical treatment.

O5-5

Time course of reverse remodeling after biventricular upgrade for conventional pacing induced left ventricular failure in congenital heart disease

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Introduction: Limited data have shown improvement of left ventricular (LV) function after upgrades to biventricular pacing (BiV) in conventional pacing induced heart failure in congenital heart disease. Exact time course and extent of reverse remodeling is, however, not known.
Methods: Nine patients with surgical (N = 8; tetralogy of Fallot = 3, ventricular septal defect = 3, other = 2) and congenital (N = 1) complete AV block who developed severe LV dysfunction after a mean of 4.9 yrs of dual-chamber RV pacing were upgraded to BiV at a median age of 6.4 yrs and subsequently followed-up for up to 1 year.

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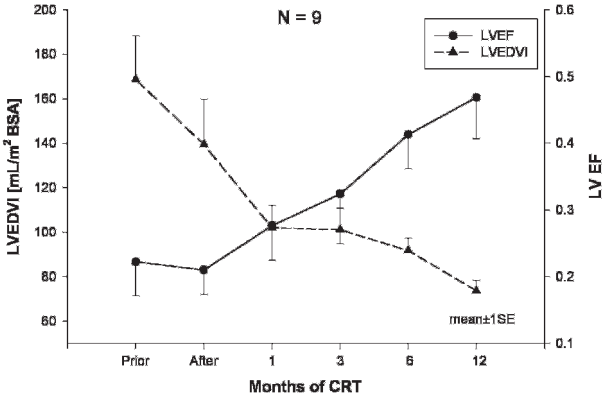
Results: Septal to posterior wall motion delay decreased from mean 269 ± 55 to 56 ± 100 ms (p = 0.003) immediately after BiV. LV enddiastolic volume index (LVEDVI) normalized after 12 months of therapy (from mean 169 ± 55 to 74 ± 11 mL/m^2 BSA, p = 0.03) with 70% of the improvement observed within the first 3 months of BiV. LV ejection fraction (EF) did not reach normal values and improvement was slower (41% of total increase at 3 months, see Fig.). All pts improved in NYHA class (from mean 2.8 ± 0.7 to 1.4 ± 0.5, p = 0.009).



Conclusions: Upgrade to BiV leads to immediate improvement in mechanical LV synchrony. This is followed by a normalization of enddiastolic volume. Improvement in EF and hence decrease in endsystolic volume is slower and does not reach normal values after one year of therapy. It remains to be seen, whether longer follow-up is be needed to observe complete reverse remodeling. (Supported by research grant of University Hospital Motol, Prague, Czech Republic, No 00064203/6301)

O5-6

Biomarker responses during long-term mechanical circulatory support in children

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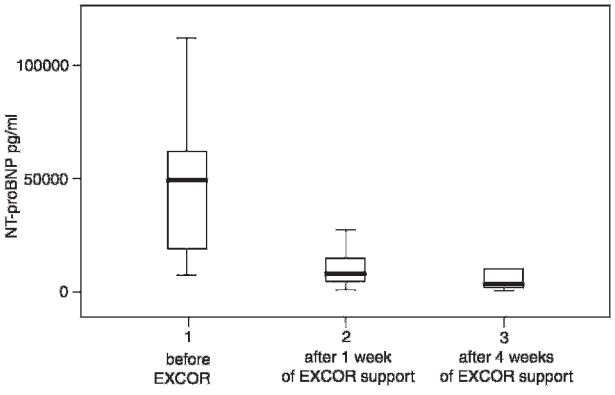
Objectives: To evaluate the effect of long-term mechanical circulatory support on the natriuretic hormone system in children and to assess whether unloading of the ventricle, haemodynamic improvement and potential myocardial recovery are associated with changes in the plasma values of natriuretic peptides.
Methods: Serial blood samples were collected from 19 children (median age 10.8, range 0.2–17.5 years) with end stage heart failure, all supported with a pulsatile ventricular assist device (Berlin Heart EXCOR). Levels of NT-proBNP were analysed before and 7 and 30 days after device implantation. In addition levels of midregion pro-ANP (MR-proANP) and BNP were determined in 13 of the 19 children.
Results: Serial measurements of NT-proBNP (Fig.), BNP and MR-proANP showed a significant decrease of plasma levels of all three natriuretic peptides within the first week of mechanical circulatory support and a further decrease between days 7 and 30. A reduction of the LVEDD, as a parameter of unloading, could be detected within the first week after EXCOR implantation. Afterwards no further decrease of LVEDD could be determined.



Conclusions: Mechanical support by a pulsatile assist device leads to a rapid unloading of the ventricle of the failing heart which can be seen echocardiographicaly in a decrease in LVEDD. Natriuretic peptides, which are highly elevated in children with end-stage heart failure, seem to be a good parameter to monitor ventricular unloading under long-term mechanical support.

O6-1

Multiplanar review of 3D Echocardiographic Datasets is an Accurate Method for Defect Sizing

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Introduction: When planning implantation of intra-cardiac devices, it is of supreme importance to select the correct device size. 3D echocardiography is emerging as a useful tool in paediatric clinical practice, and we have found the multiplanar review facility (MPR) to be useful and accurate in size determination of defects in both simple and complex congenital lesions.
Methods: Thirteen patients, aged 6 months to 46 years underwent sizing of defects by 2Dimensional echocardiography, 3D MPR, and intra-procedure measurement (balloon-sizing, angiographic measurement, or intra-operative direct measurement). The diagnoses were as follows: ventricular septal defect (n = 8), patent ductus arteriosus (n = 2), atrial septal defect (n = 1), para-prosthetic valvar leak (n = 2).
RT3DE data acquired as full volume loops were analysed offline in multiplanar review mode (MPR), using 3 orthogonal planes to determine accurately the limits of the defect. These dimensions were compared to balloon sizing in 5 patients and intra-operative sizing in 8 patients, using Spearman rank correlation to assess accuracy between methods.
Results: Correlation between 2D echocardiography and intra-procedure measurement was 0.788, and between 3D MPR and intra-procedure measurement was 0.975 (p = 0.001). In one patient with a prosthetic para-valvar leak, MPR identified discrete 2 leaks, where 2D echocardiography had only been able to identify 1.

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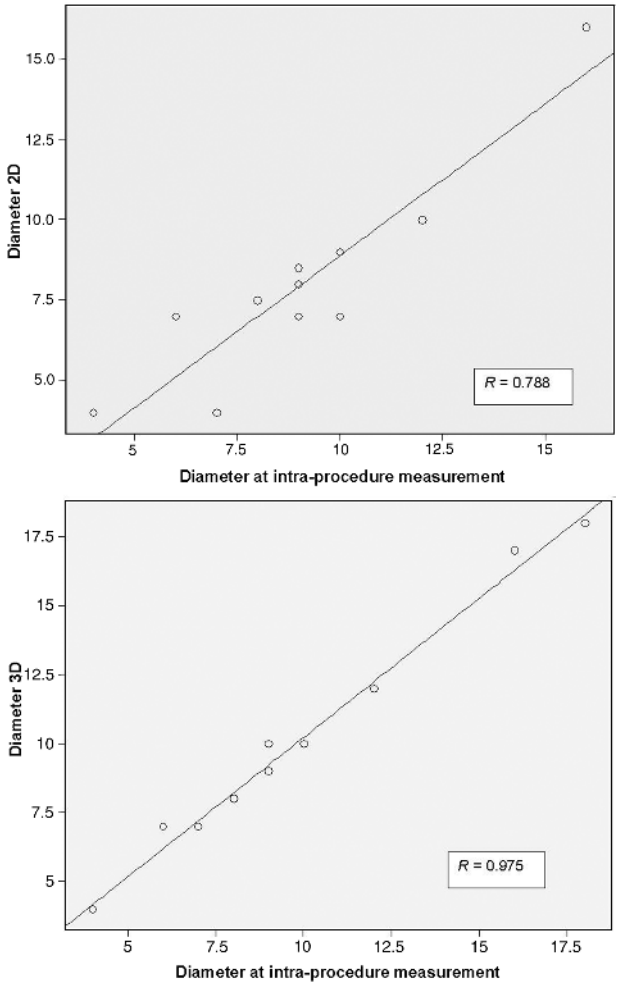

Conclusions: MPR of 3D echo data sets is a rapid and accurate method of determining size and is an extremely useful tool in preparation for interventions in complex congenital heart disease.

O6-2
A Meta-Analysis of percutaneous versus surgical closure of ostium secundum atrial septal defects

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Background: Percutaneous ASD closure has become recently available. In literature there are some observational studies in which results of these two techniques have been compared. However a comparison in a very large data set is lacking. To overcome this issue and to provide an evidence-based evaluation of the comparison between these two techniques we performed a comphehensive meta-analysis of all currently available studies comparing surgery and the transcatheter approach.
Methods: Electronic databases were systematically searched for pertinent clinical studies comparing the two methods of closure (percutaneous and surgical) published up to December 2006 and reporting on >20 patients. The primary endpoints were the occurrence of death, of total and major early complications. Pooled estimates for odds ratios (OR) were computed according to random-effect method (95% confidence intervals), with statistical inconsistency appraised with I2. Two-tailed p values and 95% confidence intervals are reported throughout, when applicable.
Results: After excluding 320 non-pertinent citations, we finally included 12 original studies (2820 patients). All studies were non-randomized comparison between percutaneous and surgical closure. No deaths were encountered with any of the two methods. Quantitative synthesis of total complications after procedure of 31% (95% CI 21–41%) in patients treated surgically while it was 6.6% (95% CI 3.9–9.2%) in subjects treated percutaneously. Comparison of percutaneous closure versus surgery showed adjusted odds ratios for total complications of 0.17 (95% CI 0.11–0.27; $p < 0.0001$).
Quantitative synthesis for major complications after procedure of 6.8% (95% CI 4–9.5%) in patients treated surgically while it was 1.9% (95% CI 0.9–2.9%) in subjects treated percutaneously. Comparison of percutaneous closure versus surgery showed adjusted odds ratios for total complications of 0.29 (95% CI 0.18–0.45; $p = 0.006$).
Conclusions: The largest cohort to date of patients with secundum atrial septal defect shows that treatemnt by a percutaneous approach has a lower rate of either total or major early post-procedural complications.

O6-3
Value of pharmacologic stress testing of so called "minimal" residual coarctation – it is time to change the traditional paradigma of good surgical results

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Introduction: Coarctation of the aorta (CoA) is one of the most common and "simple" congenital lesions. Traditionally, a residual gradient after surgical repair below 20 mmHg at rest (or during catheter evaluation) is referred as an excellent result without need for re-intervention. However, there is a growing population of patients with these residual lesions but significant problems, such as hypertension (at rest, exercise induced), pathologic 24-hr blood-pressure monitoring or left ventricular hypertrophy. We hypothesised that pharmacologic stress testing in these patients may reveal the true impact of the residual lesion and that the traditional treatment recommendation may need modification.
Methods: Prospective investigation over a 6 month period in all patients with so called minimal residual CoA and other pathologic problems (see above). After MRI assessment, catheter based pharmacologic stress protocol using bolus administration of Orciprenaline. Additional factors evaluated were the minimal diameter of the re-CoA site and the diameter of the aorta at the level of the diaphragm (dAo). Subsequent stent implantation was performed and the pharmacologic stress testing was repeated immediately thereafter.
Results: 26 patients were included. The initial CoA diameter was 9.27 mm (SD 3.19 mm) or 56% of dAo (17.50 mm; SD 4.15 mm). The gradient at rest was 17.54 mmHg (SD 7.53 mmHg), the gradient after the orciprenaline bolus increased to 61.42 mmHg

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(SD 21.60 mmHg). Stent-Implantation (CP-Stents) increased the diameter of the CoA-site to 16.35 mm (SD 3.44 mm) or 95% of dAo. The residual gradient was 0.8 mmHg (SD 2.48 mmHg) and after a second bolus of Orciprenaline 5.88 mmHg (SD 8.16 mmHg).

Conclusions: Pharmacologic stress testing by using Orciprenaline can unmask the significance of so called minimal residual CoA and is an important tool in assessing these patients. In our opinion these patients should be treated by catheter intervention (i.e. stenting to dAo diameter) to minimize the impact of this long standing disease. The paradigm of "good surgical results" needs urgent modification in the long-term assessment of these patients.

O6-4

Growth among children with hypoplastic left heart syndrome (HLHS)

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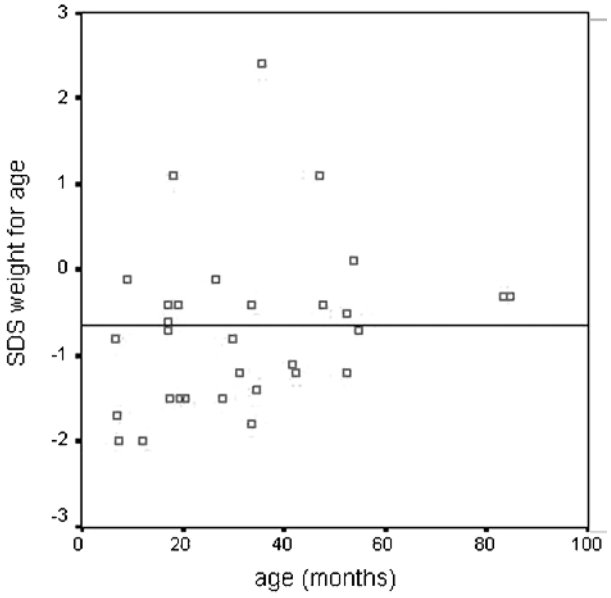
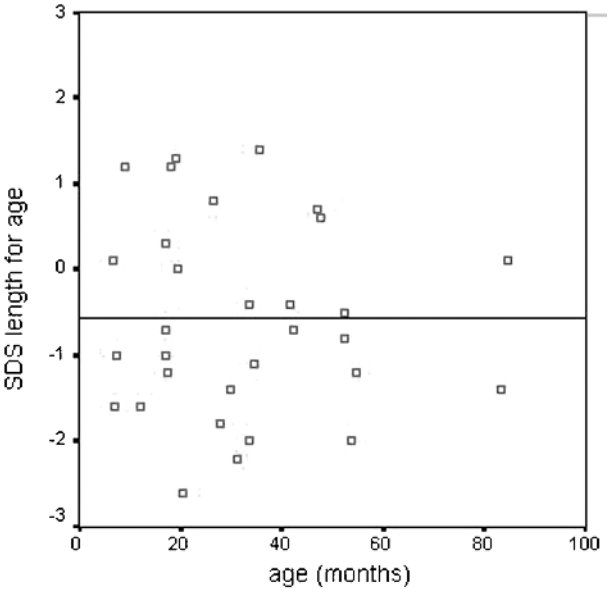
Introduction: Growth retardation is common among infants with haemodynamically important congenital heart disease. Previous studies have shown significant growth failure in 43–58% of children with HLHS after staged Norwood procedure.

Methods: Growth data of 31 consecutive patients with HLHS, survivors of Norwood operation between December 1999 and May 2006, were retrospectively analyzed. Follow-up ranged from 6.4 to 84.2 months (mean 32 months). Analyzed data were: weight at birth, weight-for-age at Glenn procedure, weight-, length- and BMI-for-age at age of one year, at Fontan operation and at last control. Measurements were expressed as standard deviation scores (SDS), using national growth charts (TNO 1997). Paired sample t-tests and independent t-tests were used for statistical analysis.

Results: Weight at birth was normal for gestational age. Norwood operation was performed at 1.5 ± 067 weeks (11 B-T shunt, 20 Sano-shunt). At Glenn procedure (mean 4.4 months) weight-for-age was −1.21 ± 0.79 SDS (n = 31). No difference was seen between Sano- and B-T-shunt. At the age of one year weight-for-age was −0.77 ± 0.95 SDS, length-for-age −0.34 ± 1.16 SDS and BMI-for-age was −0.82 ± 0.94 SDS (n = 27). At Fontan operation (mean 34.8 months) weight-for-age was −0.33 ± 1.07 SDS, length-for-age −0.38 ± 1.26 SDS and BMI-for-age −0.24 ± 0.91 SDS (n = 12). At last control weight-for-age was −0.69 ± 0.96 SDS with no patients under -2. Length-for-age was −0.58 ± 1.12 SDS with 2 patients under -2 (figure). BMI-for-age was −0.55 ± 1−02 SDS. In 38% mild underweight was seen (SDS BMI −1.0 to −2.0) and 6,4% had severe underweight (SDS BMI <−2.0). The SDS weight-for-age at birth was significantly higher than at time of Glenn operation (p = 0.000). At the age of one year SDS weight-for-age was significantly higher than at time of the Glenn operation (p = 0.005). The improvement of SDS weight-for-age and length-for-age between one year and Fontan operation did not reach significance (p = 0.254, resp. p = 0.627).

Conclusion: Growth failure is most obvious before Glenn-procedure. After Glennoperation improvement of growth is seen. Our patient group shows better growth than patient groups described in international literature. One of the reasons may be the special attention that was given to feeding and growth during frequent control visits.





O6-5

Impact of early and aggressive treatment with amiodarone on the therapeutic success and outcome in patients with postoperative tachyarrhythmias

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Introduction: Tachyarrhythmias in the early postoperative setting after repair of congenital heart defects have significant impact on the perioperative morbidity and mortality. Thus adequate rhythm and heart rate control is an important tool in the management of these patients.

Methods: Prospective data collection over an 8 year period in all patients admitted to a tertiary paediatric cardiac ICU who received intravenous Amiodarone for postoperative arrhythmias. Patients with preoperative treatment were excluded as well as patients with treatment for resuscitation purposes. Parameters monitored were the cumulative dose of Amiodarone, the total dose until rate

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and rhythm control was achieved, the time until treatment was started after presentation of the arrhythmia, the total treatment time and length of stay in the PCICU. Finally, the impact of a new treatment protocol was assessed.
Results: 71 children could be identified; in 40 patients (treatment group A) therapy was started more than 60 min after the first presentation of the arrhythmia, in 31 patients (treatment group B) therapy started within 60 min. Heart rate control was achieved in 200 min (A) and 60 min (B), rhythm control in 600 min (A) and 150 min (B) [mean values]. The mean cumulative dose of Amiodarone for rate control was 6.1 mg/kg (A) and 3.8 mg/kg (B). The total time for intravenous therapy was 56.6 hrs (A) and 45.7 hrs (B). Length of stay in the PCICU was 5.6 days (A) and 3.6 days (B). After implementation of the new treatment protocol the majority of patients was treated within 60 min of the presentation of the first arrhythmias. The treatment protocol developed consisted in a stepwise bolus administration of repetitive doses of 5 mg/kg over 1 hr followed by a continuous administration of 10–20 mg/kg/day and was well tolerated.
Conclusions: Early and aggressive therapy of postoperative tachyarrhythmias by using a specific Amiodarone treatment protocol has significant impact on perioperative morbidity, treatment time and cumulative dose applied and may reduce LOS in the PCICU.

O6-6
Remote Magnetic Navigation: A new approach for mapping and ablation of supraventricular tachycardias in children and patients with congenital heart disease

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Introduction: Remote magnetic navigation offers a new approach for mapping and ablation of different arrhythmia substrates. Advantages of the system might include increased catheter stability, maneuverabilty and reduced fluoroscopy time. First reports in adults show promising results, but no sufficient data for children or patients with congenital heart disease are available yet.
Methods: The data of the electrophysiological studies using the remote magnetic navigation system (Niobe®, Stereotaxis Inc.; Siemens) of our first 9 patients (pts) were reviewed. Congenital heart disease was present in 5/9 pts. (Table 1). In 8/9 pts an additional 3D mapping system (4x Carto®, 3x Carto-Merge®, 1x NavX®) was used. All pts had supraventicular tachycardia (SVT) including intraatrial reentrant tachycardia (IART; n = 4), focal atrial tachycardia (FAT; n = 3) or tachycardia using an accessory pathway (n = 2). Mapping and ablation was performed using the currently available 7F non-irrigated 4mm tip catheter (Navistar RMT® n = 7, Helios® n = 2, both Biosense Webster).
Results: Integration of all 3D-mapping systems was possible without problems. We observed exact and reliable steering of the catheter in the right (RA) and left atrium (LA). Mapping of the tachycardia substrate was possible in all pts. In 4/9 pts mapping of the RA, in 1 pt mapping of the LA and in 4/9 pts biatrial mapping was performed. LA access was achieved by transseptal puncture (n = 1), via an atrial septal defect (n = 1) or retrogradely via the aortic and mitral valve (n = 2). Using the magnetic catheter, 2/4 tachycardias originating from the RA (Pt. Number 1 and 8) were successfully ablated. No procedure-related complications occurred.

Patient Number	Mapping	3D System	Fluoroscopy time in min (including ablation)
1	RA, RAA	Carto	4.2
2	RA, Coronary Sinus	NavX	5.8
3	LA, LAA (transeptal)	Carto	16.4
4	RA	Carto	3.9
5	RA	no	5.1
6	RA, LA (retrograde)	Carto-Merge	19.3
7	RA, LA (retrograde)	Carto-Merge	49
8	RA	Carto	35
9	RA, LA (via ASD)	Carto	15.4

Patient Number	Age in years	Congenital Heart Disease	Arrhythmia
1	5	No	Focal atrial Tachycardia (FAT) originating in Right atrial appendage (RAA)
2	7	No	Permanent Junctional Reentry Tachycardia
3	12	No	FAT originating in Left atrial appendage (LAA)
4	13	No	FAT from TV-anulus
5	11	Ebstein's Anomaly	Wolff Parkinson White Syndrome
6	23	Transposition of the Great Arteries, Senning operation	IART
7	50	Double Inlet Left Ventricle, Fontan Operation	IART, Atrial Fibrillation
8	55	Atrial Septal defect, Pulmonary Hypertension	IART
9	66	Atrial Septal Defect	IART

Conclusions: By providing exact and reliable catheter steering, remote magnetic navigation allows mapping of different SVT substrates in patients with small cardiac chambers or complex anatomy. Reduction of fluoroscopy time seems possible. Catheter ablation can be perfomed in selected subgroups. The development of irrigated tip catheters should enable safe and reliable catheter ablation in the majority of cases.

O7-1
The utility of ECG criteria in Pediatric ARVC

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The electrocardiogram (ECG), when appropriately analyzed, is recently reported to have high sensitivity (SENS) and high specificity (SPEC) for arrhythmogenic right ventricular cardiomyopathy (ARVC) within adults. We sought to identify the SENS and SPEC of similarly analyzed ECGs within children.
Methods: Children presenting with LBBB VT or a positive family history of ARVC underwent noninvasive assessment for Task Force criteria for ARVC, including ECG. Those identified with an additional ARVC criterion on non-invasive assessment underwent invasive assessment including angiography and biopsy. ECGs from both presentation and latest follow-up were analyzed using the

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computer-derived "measurement matrix" from a GE Medical ECG machine for QRS duration, the ratio of QRS duration in V1+ V2+V3/V4+V5+V6, and S wave durations in V1, V2 and V3. T and T´ wave amplitudes and maximum negative T wave amplitude were measured in in V1, V2 and V3. All measurements except the QRS duration ratio were analyzed using regression equations derived from a group of 139 normal school children aged 5–15 years. This normal group was also used to assess specificity of individual ECG criteria.
Results: ECGs were available in 34 children who met Task Force criteria for ARVC aged 4.0 to 17.6 (12.5 ± 3.6) years at presentation and 8.4 to 18.2 (16.3 ± 2.3) years at follow-up. Normalized QRS duration had a SENS of 8/34 (24%) and SPEC of 135/139 (97%) for ARVC at presentation and SENS of 11/33 (33%) at follow-up; only 2/34 patients had a QRSD ≥ 110 at presentation. QRS duration ratio was unable to discriminate between normals and ARVC. Lead V2 provided the best discrimination by S wave duration with a SENS of 11/34 (32%) and SPEC of 132/138 (96%) for ARVC at presentation and SENS of 12/33 (36%) at follow-up. Normalized minimum T amplitude had a SENS of 15/34 (44%) and SPEC of 135/139 (97%) for ARVC at presentation and SENS of 16/33 (48%) at follow-up. Only 11/34 (32%) children met adult T-wave criteria.
Conclusion: Pediatric ECG criteria have moderate sensitivity for ARVC when normalized for age. Normalized ECG criteria can identify additional children with ARVC who do not meet adult ECG criteria.

O7-2
Predictors of Response to Cardiac Resynchronization Therapy (CRT) in Pediatric and Congenital Heart Disease. Subanalysis of a Retrospective European Multicenter Study

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Introduction: Response to cardiac resynchronization therapy (CRT) in a heterogeneous population with pediatric and congenital heart disease may differ from adult ischemic and idiopathic cardiomyopathy.
Methods: Retrospectively collected data from 109 patients aged 0.24–73.8 (median 16.9) yrs with congenital heart disease (N = 87), cardiomyopathy (N = 12), congenital complete AV block (N = 10) with systemic left (N = 69), right (N = 36) or single (N = 4) ventricular dysfunction and spontaneous (N = 25) or pacing induced (N = 84) ventricular desynchronization subjected to CRT and followed-up for a median of 7.5 mo were analyzed. Predictors of clinical response and reverse remodeling were evaluated.
Results: 16.1% of patients were identified as non-responders. Predictors of non-response were the presence of primary cardiomyopathy (univariate: 40.0% vs 3.8%, $p < 0.001$; multivariate: $p < 0.001$), higher NYHA class (univariate: median 3.5 vs 2.0, $p < 0.001$; multivariate: $p = 0.004$) and greater systemic ventricular enddiastolic dimension (univariate: median z-score: +6.36 vs +2.66, $p = 0.003$; multivariate: NS). Major improvement in systolic function (over the 50th percentile of the study group) was predicted by the presence of systemic LV (univariate: 95.0% vs 47.4%, $p = 0.001$; multivariate: $p < 0.001$), lower initial ejection fraction (univariate: median 25 vs 30%, $p = 0.062$; multivariate: $p < 0.001$) and age at CRT (univariate: median 11.0 vs 17.9 yrs, $p = 0.032$; multivariate: NS). Increase in ejection fraction/fractional area of change was better for systemic LV than RV (mean +15 vs +6%, $p = 0.029$).
Conclusions: Primary dilated cardiomyopathy and high initial NYHA class were two independent predictors of poor response to CRT. Improvement in systolic function was significantly better in patients with systemic LV than RV. Other factors, like QRS duration and change, age or presence of pacing-induced vs spontaneous ventricular desynchronization did not play a role in prediction of outcome. (Roman A. Gebauer was supported by the Research Project No 00064203/6301 of University Hospital Motol, Prague).

O7-3
Lack to Prove Hemodynamic Short-Term Benefit of Biventricular Pacing in Patients with d-TGA after Atrial Switch

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Aim of the study: To prove hemodynamic benefit of ventricular resynchronisation (CRT) in patients with d-TGA and atrial switch (Mustard or Senning).
Introduction: Patients with d-TGA after atrial switch frequently have enlarged morphological right (systemic) ventricles (mRV), wide QRS and low heartrate. CRT should improve effectiveness of myocardial work and is shown to be beneficial in a long term application in some patients.
Patients and Methods: 8 Patients with d-TGA, palliated with atrial switch, were investigated. All patients were male, aged 19.6 to 29.7 years and were clinically stable (NYHA 1). Three patients had a permanent pacemaker (DDD) with the ventricular leads placed in the mRV.
Under general anaesthesia residual lesions like buffle leaks, obstruction of inflow or outflow or valve incompetence was excluded by transesophageal echocardiography and complete invasive evaluation. Transvascular pacemaker-leads were placed in the right atrium and the free wall of the mRV and the morphological left ventricle (mLV). In addition, a Swan-Ganz-catheter was placed.

If a permanent pacemaker was in place it was disabled during the evaluation. Cardiac output was measured by the principle of Fick and indexed to body surface (CI). Measurement was done under spontaneous rhythm and pacing with a frequency of 80 bpm in the following modes (ventricular stimulation given in brackets): A00, DDD (mRV), DDD (mLV), DDD (biventricular).
Results: Mean spontaneous heartrate was 56 (42–68) with a mean CI of 2.6 (1.7–3.7). With pacing the following values were obtained:
DDD (mRV) 2.6 (1.7–3.9), DDD (mLV) 2.6 (1.7–3.7), DDD (biventricular) 2.7 (1.5–4.2). A00- or DDD-pacing improved CI in two patients. There was no benefit of stimulation in the mRV or biventricular, compared to A00 or mLV-stimulation. There was no significant change in filling- or arterial pressures.
Conclusion: CRT shows no short time benefit in patients with d-TGA after atrial switch. Invasive testing is not appropriate to identify patients that might have a benefit of CRT. Further studies are requested to develop parameters for identification of patients in whom CRT is beneficial.

O7-4
Implantable cardioverter defibrillator therapy in children: long term clinical outcome and risk factors for shocks
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Introduction: In recent years implantable cardioverter defibrillator (ICD) therapy is used more often in children at high risk for life-threatening arrhythmias. However, experience of ICD therapy in children is still limited and random. The purpose of this multicenter study is to identify predictors for ICD therapy and to analyze outcome in pediatric ICD recipients.
Methods: From 1995 to 2006, 45 pts in The Netherlands <18 yrs received an ICD. Since 2002 all pts are prospectively followed every year. Mean age at implant was 10.8 ± 5.2 yrs. Both epicardial/subcutaneous (10 pts) and transvenous approaches (35 pts) were used. Indications were SCD survivors (n = 23), syncope/VT (n = 10), prophylaxis/familial SCD (n = 12). Underlying cardiac disorders were primary electrical disease (55%), cardiomyopathy (20%), congenital heart disease (17%), others (8%).
Results: Mean follow-up was 44 ± 32.9 months. Three pts (7%) died and 1 pt (2%) underwent HTX. ICD related complications occurred in 6 pts (13%), including lead dislocation (1), lead fracture (2) and DFT rise (3). 14 pts (31%) received appropriate shocks, median 6.5 shocks per pt; 12 pts (27%) received inappropriate shocks, median 3.5 shocks per pt. Appropriate shock therapy was similar in the group of SCD survivor (43%) and syncope/VT group (44%) (n.s.). In these patients, the group of ICD implantation under 12 yrs received significantly more appropriate shocks than the older age group (12–18 yrs), 12/22 pts (55%) vs. 2/23 pts (9%) respectively (p = 0.003). However, pts with ICD therapy as prophylaxis/familial SCD (n = 12) received no appropriate shocks during mean FU of 44.4 ± 25.7 months (p = 0.03). There were no significant differences in inappropriate shock therapy between different age groups or between different cardiac disorders.
Conclusions: In a pediatric population ICD therapy for ventricular arrhythmia in children under 12 years of age is associated with much higher incidence of appropriate shocks than ICD therapy for ventricular arrhythmia in older children. The benefit of prophylactic ICD therapy in the absence of life threatening arrhythmia could not be demonstrated and remains to be determined.

O7-5
Cryoablation at growing myocardium: Results of intracoronary ultrasound and coronary angiography after energy application
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Introduction: Animal studies and clinical observations have demonstrated that radiofrequency current application at growing myocardium may result in coronary artery obstruction. Recently, cryoenergy has emerged as an effective alternative to radiofrequency ablation of arrhythmogenic substrates in pediatric patients. Up to now, there is a lack of experimental data concerning the effects of cryoenergy application at growing myocardium.
Methods: During general anesthesia selective coronary angiography of the right and left coronary artery was performed in 10 piglets (age 6 weeks, body weight 14–18 kg). Subsequently, cryoenergy (Freezor Xtra®, 7F, 6 mm tip electrode) was delivered at −75°C for 4 minutes at the atrial aspect of the tricuspid valve annulus in a posterior and lateral position. Additional cryoenergy lesions were induced via a retrograde approach at the lateral and posterior atrial and ventricular aspect of the mitral valve annulus, respectively. Five animals were restudied after 48 hours by coronary angiography and intracoronary ultrasound (Jovus Avanar F/X, 2.9 F) and in the remaining 5 piglets after 3 and 6 months, respectively.
Results: Selective coronary angiography did not demonstrate any evidence for coronary artery obstruction after 48 hours, 3 and 6 months after cryoenergy application. In addition, intracoronary ultrasound studies did not reveal any intima plaque formation or stenosis, respectively.
Conclusion: By the means of the present study, we did not observe any affection of the coronary arteries after cryoenergy application at growing myocardium in young piglets.

O7-6
Catecholaminergic Polymorphic Ventricular Tachycardia in Pediatrics
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Background: Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare diagnosis. Patients present with a history of exercise induced syncope (ES) or aborted sudden cardiac death (SCD). Little pediatric data concerning this arrhythmia exists. The purpose of this study was to illustrate the diagnosis, family history and outcomes of a group of pediatric patients with a diagnosis of CPVT.
Methods: An IRB approved, retrospective search of 5 pediatric cardiology databases was performed to identify patients. Inclusion criteria included: age <18 at diagnosis and documented CPVT. Data collected included: age, age at diagnosis, presentation, family history, exercise stress test (EST) and ECG results, defibrillator

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implantation (ICD), shocks, and medications. Patients were further identified as index or non-index. Index patients were defined as the first patient diagnosed with CPVT. Results: Fourteen patients (7 male/7 female) with an average age of 10.8 years at diagnosis met inclusion criteria. Seventy five percent of the patients were the index case and all have been followed for an average of 2.3 years (range 1–11). Presenting symptoms included ES in 65%, SCD in 14% and family history in 21%. Sinus bradycardia was the only diagnosis on 36% of ECGs. During EST, 78% had polymorphic ventricular tachycardia, 14% had ventricular couplets and 8% did not have EST due to age. 92% of patients had ICD implants with 3 appropriate and 21 inappropriate discharges. All patients are on medications (Nadolol 13, Atenolol 1). Family screening identified 13 additional patients with CPVT. Genetic screening has resulted in 2 positive tests. Conclusions: CPVT should be considered in all patient with syncope or sudden cardiac death during exercise. Performing an EST appears to be an effective method of diagnosing CPVT. Family screening should be performed. ICD implants, although recommended, often resulted in inappropriate shocks.

O8-1
Immediate haemodynamic effect and short term clinical follow up following percutaneous pulmonary valve implantation

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Introduction: Percutaneous pulmonary valve implantation (PPVI) was introduced in 2000 as a new treatment of right ventricular outflow tract (RVOT) failure and is on the verge of becoming clinical routine.
We report on the immediate effects of PPVI on the outflow tract gradient and pulmonary valve competence, as well short term changes in symptoms and exercise capacity.
Methods: All patients undergoing PPVI in our institutions were included in the study. Invasive haemodynamic measurements were made immediately before and after valve implantation. Magnetic resonance (MR) imaging was performed prior to and within 1 month after the procedure and New York Heart Association (NYHA) functional class recorded.
Results: Between September 2000 and December 2006, 144 patients (median age 21.2 years, range 7–71 years; 59 female) underwent PPVI, by a single operator, at our institutions. Patients with pulmonary atresia/VSD and tetralogy of Fallot comprised the majority (56%) and 114 patients (78%) had existing homograft conduits. 61% had predominantly stenotic RVOT pathology (as defined by an invasive ventricular-pulmonary artery gradient of ^{3}30 mmHg) and the remainder predominantly regurgitation.
Following PPVI outflow tract gradient fell from 50.5 ± 16.6 to 20.5 ± 10.9 mmHg ($P < 0.0001$).
No patient in either group had angiographically significant pulmonary regurgitation after PPVI. MR assessment showed an increase in indexed right ventricular effective stroke volume from 39.5 ± 10.6 ml/m^2 to 43.8 ± 9.0 ml/m^2 ($P = 0.0001$) and a reduction in pulmonary regurgitant fraction from 19.8 ± 14.7 to 2.9 ± 4.6% ($P = 0.0001$).
One month following PPVI patients reported to be less symptomatic and mean NYHA class had fallen from 2.3 to 1.4 ($P < 0.0001$).
Conclusion: PPVI resulted in a significant fall in ventricular-pulmonary artery gradient in patients with RVOT stenosis and the implanted valve was competent in all. One month after PPVI patients were less symptomatic and effective cardiac output at rest was increased.

O8-2
Medium term results of stent implantation for pulmonary artery branch stenosis in infants and small children

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Introduction: Stenting of stenotic branch pulmonary arteries (PA) in young children is controversial because of potential negative effects on PA growth and risk of restenosis. The objective of this study is to evaluate clinical outcome of stent implantation for PA branch stenosis (PABS) in young children.
Methods: We retrospectively analyzed data of 42 stent implantations for postoperative PABS in 33 consecutive children under 15 kg.
Results: Diagnosis included truncus arteriosus, TGA, hypoplastic left heart syndrome, tetralogy of Fallot and pulmonary atresia-VSD-MAPCA. Stents were implanted percutaneously ($n = 23$ pts) or intraoperatively as part of a hybrid approach ($n = 10$ pts). Age at implant was 22.4 ± 20.1 months, weight 9 ± 3.9 kg, median follow-up was 2.2 yrs ranging from 0.1 to 10.4 yrs. Stent diameters ranged from 4 to 10 mm, lengths ranged from 8 to 19 mm. Stent dislodgement/malposition occurred in 3 pts (9%), there were no procedural deaths or other complications. Acute results in pts with biventricular hearts showed decrease of RV pressure from 64 ± 16 to 48 ± 10 mmHg ($P < 0.05$), decrease of RV/LV pressure ratio from 0.75 ± 0.20 to 0.56 ± 0.14 ($P < 0.05$) and decrease of PA pressure gradients from 45 ± 22 to 27 ± 22 mmHg ($P < 0.05$). Perfusion improved in unilateral PPS from 25 ± 9% to 37 ± 8% ($P < 0.05$). Two pts with pulmonary atresia-VSD-MAPCA died during FU. Re-stenosis occurred in 42% ($n = 14$) requiring catheter ($n = 8$) or surgical ($n = 2$) re-intervention in 30% ($n = 10$). Patients with stents implanted <1 yr had higher rate of restenosis than pts with stent implanted >1 yr, 54% versus 36% ($P < 0.05$). Post-stent diameters of the branch PA increased from 5.3 ± 1.8 mm to 7.5 ± 3.3 mm ($P < 0.05$). At last follow-up mean RV pressure was 46 ± 15 mmHg and mean PA gradient was 36 ± 14 mmHg.
Conclusions: Medium term results of stent implantation for PABS in infants and small children are encouraging. PA growth remains preserved. However, reinterventions are often required due to the high incidence of restenosis .

O8-3
ASD and PFO closure with the Solysafe device. First clinical experience

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Objectives: We report the results of a prospective multicenter pilot study performed in Germany, Sweden and Switzerland with a new

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self-centering device for transcatheter closure of an ASD or a PFO called the Solysafe Septal Occluder.
Methods: The Solysafe Septal Occluder (Swissimplant AG, Solothurn, Switzerland) has been developed for transcatheter closure of ASD and PFO. It consists of a self-centering device with two foldable polyester patches which are attached to eight metal wires made of Phynox, a cobalt-based alloy used in surgical implants for years (Fig. 1). Two wire-holders keep the wires fixed at the proximal (right-atrial) and distal (left-atrial) end of the device.
Results: In 44 patients the device was successfully implanted. In 15 patients with a median age of 40 years (range 6–76 years), a Solysafe device was successfully implanted in an ASD. The median size of the stretched defects was 17 mm (range 10–21 mm). Three 15 mm devices have been used, eight 20 mm devices and four 25 mm devices. Procedure time ranged from 40 min to 107 min (median 66 min). Fluoroscopic time ranged from 5.3 min to 17.5 min (median 12 min). In 29 patients with a median age of 47 years (range 15–78 years), a Solysafe device was implanted in a PFO. The procedure time ranged from 21 to 155 min (median 51 min). Fluoroscopic time ranged from 3.1 min to 31.3 min (median 7.6 min). At discharge, 1 of the 29 patients (3%) had a small shunt. No patient neither in the ASD-group nor in the PFO group had any major complication.
Six months after implantation, the overall closure rate with the Solysafe septal occluder in both the ASD and the PFO group is 100% (44/44).
Conclusion: With the self-centering Solysafe Septal Occluder, PFO and ASD with a stretched diameter up to 21 mm can effectively be closed with very high occlusion rates.

O8-4
Biocompatibility of atrial septal defect closure devices: Immunohistochemical characterisation of neo-tissues
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Objective: Safety and efficacy of interventional treatment of septel defects has been studied thoroughly. In contrast, little attention has been paid on biocompatibility. In our study, we are presenting data from serial immunohistochemical examinations of human explanted septel occlusion devices.
Materials and methods: ASD or VSD occlusion devices (Amplatzer n = 7; Cardioseal/Starflex n = 3) were processed using a uniform protocol after surgical removal from humans (implantation time 5 days to 4 years). Devices were fixed in formalin and embedded in methylmethacrylate. Serial sections were obtained by sectioning with a hard sword, or with a diamond cutter and grinding, thus saving the metal/tissue interface for histological evaluation. Immunohistochemical stainings were performed using a standard protocol.
Results: Superficial endothelial cells with positive staining for von Willebrand factor (vWF) were seen in all specimen. Even in the implant with the shortest implantation time (5 days; Amplatzer ASD occluder) there was some positive staining of the superficial cell layer indicating early adherence of endothelial cells. Neo-tissue formed within the implants after initial fibrin condensation showed no staining for marker of mature muscle cells (Caldesmon, Myosin, Desmin), whereas a more unspecific marker (Smooth muscle actin) stained positive in single specimen. Good vascularisation of the neo-tissue was demonstrated with the typical immunohistochemical pattern of small vessels. In single implants, characterisation of inflammatory cells was achieved, indicating some lymphcellular inflammation with mixed B and T cells beside a histiocytic foreign body reaction.
Conclusion: This is the first presentation of results from serial immunohistochemical stainings of a collection of human explanted septal occlusion devices. Our results indicate that endothelialisation may occur earlier than assumed from morphological studies. Poorly differentiated fibromuscular cells could be identified within the implants after cellular organisation of the initially formed thrombotic material.

O8-5
Stent implantation in the arterial duct from the axillary artery in duct dependent pulmonary blood flow: anatomical and technical aspects
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Introduction: The arterial duct in patients with pulmonary atresia or complex pulmonary stenosis often describes a tortuous course and originates more from the transverse arch than in left heart obstructive lesions. Stent implantation in these ducts as an alternative to a systemic to pulmonary artery shunt from the femoral artery is often impossible. We describe a series of patients where the duct was addressed from the axillary artery; anatomical and technical aspects are discussed.
Patients and Methods: From 6/96–1/07, 53 newborns with duct dependend pulmonary blood flow were treated by stent implantation of the arterial duct. 8 of them were addressed by axillary access: male (n = 7), female (n = 1), (bodyweight: 2.6–3.9 kg (m = 3.26 kg)). Diagnoses: Pat/IVS (n = 2), Pat/VSD (n = 2), Pat/UVH (n = 2), TOF (n = 1), PS/DORV/TGA (n = 1). Dextrocardia was present in 2 pts. a bilateral duct in one.
One pt. needed cut down of the axilla, the others were punctured percutaneously. No introducer sheaths larger than 4–5F were used. In 7 pts premounted coronary stents were used, in 2 pts. 2 stents were used to cover the duct completely. The stent length varied between 10 and 24mm, the final width from 3.5 to 4.5mm.
Results: Entering the axillary artery was possible in all. Stent implantation was successful in all but one, where a dislodged stent was removed surgically. In 6 pts the left and in 2 pts the right axillary artery was engaged. All patients could be stabilized by the intervention besides one. Two pts died despite successful intervention; one due to cardiac shock by admission, another one with bilateral ducts because of pulmonary run off. Re-dilation/second stent were necessary in 2 patients, two pts got a surgical shunt. Based on the underlying heart defect 2 pts had univentricular and one biventricular repair, 3 are awaiting surgery.
Conclusion: Axillary access should be considered in newborns scheduled for stent implantation in the arterial duct. The technique of duct stenting is improved by using retrievable stents and innovative stent and balloon technology. Favourable anatomy for stent implantation and saving femoral artery perfusion are the main issues for this approach.

O8-6
Outcome of Patent Ductus Arteriosus Stenting in Patients with Ductal Dependent Pulmonary Circulation
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Objective: We sought to assess the outcome of transcatheter ductus arteriosus stenting in newborns with ductal dependent pulmonary circulation.
Background: Better results of ductal stenting have been reported recently. These results encouraged us to offer this palliation to our patients.

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Methods: Twenty one patients with ductal dependent pulmonary circulation were brought to the catheterisation laboratory for ductal stenting. Five patients did not qualify because of complex tortuosity of the ductus arteriosus or branch pulmonary artery stenosis. Stent implantation was attempted in the remaining sixteen patients. The duct was crossed with a 0.014-inch wire. A low profile pre-mounted coronary stent was implanted in the duct without using a long delivery sheath. Attempts were made to cover the entire length of the ductus arteriosus.
Results: The mean age of the patients at the time of ductal stent implantation was 24 ± 17.5 days. The mean body weight was 2.9 ± 0.35 kg. The morphology of the ductus was, mild tortuous in 4, moderately tortuous in 4, conical in 4, and vertical in 4 patients.
Ductal stenting was successful in 14 patients. The mean diameter of the ductus arteriosus was 3.9 ± 0.5 mm, mean diameter and length of the stent implanted were 3.9 ± 0.72 mm and 15.4 ± 3.16 mm respectively. Mean fluoroscopy and procedure times were 22.16 ± 12.5 minutes and 107.9 ± 34.5 minutes. The ductus arteriosus was not completely covered with stent in 5 patients at the time of primary procedure, 4 of them required re-stenting for significant desaturation, of these, one died despite successful stenting. Another patient died due to aspiration pneumonia. Of the 12 survivors, five underwent Glenn shunt with two mortalities related to pulmonary hypertension, one has undergone biventricular repair and the remaining six are doing well with a mean oxygen saturation of 85% at a mean follow-up 13 ± 6 months.
Conclusion: Stenting of ductus arteriosus, including moderately tortuous ducts is a safe palliation for patients with ductal dependent pulmonary circulation. Incomplete stenting of the duct invariably results in ductal stenosis and compromised pulmonary flow.

O9-1
Open heart surgery in neonates and premature infants weighing less than 2.5 kg
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Background: Low birth weight infants may require early surgical treatment of congenital heart defect because of their poor clinical status. Early surgical repair has been shown to be preferable to medical management or palliative surgery with delayed definitive repair.
Methods: From November 1997 to December 2006, 46 consecutive neonates weighing less than 2500 g underwent cardiopulmonary bypass operations for complete correction of congenital heart defects (n = 34) or Norwood stage I palliation for hypoplastic left heart syndrome (n = 12). A retrospective analysis was performed to evaluate early and midterm outcome. The study group included 23 males and 23 females with a median age of 10 days (2 to 110 days) and a median weight of 2260 g (1280 g to 2480 g). 34 children (74%) were born prematurely. 7 patients were critically ill and 21 were ventilated preoperatively. Diagnoses included transposition of the great arteries (13), hypoplastic left heart syndrome (12), ventricular septal defect with interrupted or hypoplastic aortic arch (5), VSD (4), tetralogy of Fallot (3), total anomalous pulmonary venous connection (2), aorticopulmonary window (1), critical valvular aortic stenosis (1), complete AV canal (1), pulmonary atresia with intact ventricular septum (1), common arterial trunk (1), left atrial tumor (1), thrombotic formation on a ventricular-atrial shunt with ASD (1).
Results: 30-day mortality was 13% (6/46). Age, gender, prematurity, additional extracardiac malformations, bypass time and aortic cross clamp time did not influence 30-day mortality. Duration of mechanical ventilation, median time at intensive care unit and median stay until discharge were 7, 12 and 26 days, respectively. At a median follow-up time of 32 months overall mortality was 19.6% (9/46). 12/37 (32.4%) survivors needed 19 reoperations and 6/37 (16.2%) patients needed interventional heart catheterization.
Conclusion: Open heart surgery can be performed in low weight infants with reduced, but acceptable early and mid-term survival.

O9-2
The Effect of Ductal Diameter on Surgical and Medical Closure of Patent Ductus Arteriosus in Preterm Neonates: Size Matters
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Objectives: To display the effect of patent ductus arteriosus (PDA) diameter on the rate of treatment success in premature neonates.
Methods: Among 537 consecutive neonates born between January 1985 and December 2005 with a diagnosed PDA, 201 premature infants (<35 weeks) treated for a hemodynamically significant PDA fitted the selection criteria and were retrospectively reviewed. Two groups were defined: group MED containing 154 (77%) babies treated successfully with cyclo-oxygenase inhibitors (Ibuprofen or Indomethacin) and group FAIL with 47 (23%) babies where medication failed to reduce the PDA diameter to hemodynamical insignificance.
Results: After unsuccessful medical treatment, 33 (70%) required surgical closure of PDA, 12 (26%) died before further treatment was possible, and 2 (4%) were discharged from hospital without clinical symptoms but still having an open PDA. PDA diameter and respiration parameters (CPAP or mechanical ventilation) were compared between the two groups: The PDA diameter in group MED (2.48 mm [range 1.3–4.2 mm]) was significantly smaller ($p < 0.01$) than in group FAIL (2.84 mm [range 1.5–5 mm]). The respiration time before PDA closure was shorter in group MED (9 days [range 0–40]) than in group FAIL (20d [range 1–59]) ($p < 0.001$). There was no significant difference of respiration time after PDA closure between the groups: group MED 12 days [range 0–60] and group FAIL 12 days [range 1–59].
Conclusion: The extended respiration time before PDA closure of preterm babies who eventually go on to surgery reflects the delay while attempting medical treatment. Fortunately, respiration time after closure shows no difference between the two groups, showing the beneficial effect of shunt elimination on lung function. Medical treatment is likely to fail with larger PDA diameters, with a cut-off at 3.5 mm. To spare unwarranted lengthy respiration and corresponding hospital stay, we recommend a strategy towards direct surgical PDA closure in preterm babies when the ductal diameter is >3.5 mm.

O9-3
Performing Ross earlier – a chance to avoid autograft dilatation and valve failure in the follow-up?
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Background: Autograft potential to grow is a major advantage of Ross procedure over other operative methods of aortic valve. However autograft valve dysfunction may lead to reoperation as a result of autograft root dilatation. It hasn't been established yet if performing Ross procedure in younger patients eliminates autograft failure due to its dilatation.
Material and methods: 68 pts who underwent Ross or Konno-Ross procedure between 1995 and 2005 were divided into 2 groups: below 7 yrs of age (A) and older (B). 30 pts were in long-term follow-up. The mean age of group A was 3.28 ± 2.7 yr and the mean follow-up was 3 ± 2.5yr. There were 6 infants in this series and the youngest patient was 6 months of age. The mean age of group B was 14.7 ± 3 yr and the mean follow-up was 3.8 ± 2 yr. There were 16 pts at age 8–14 yrs and 29 pts above 14 yrs. Autograft annulus size, autograft sinus diameter and valve insufficiency (AI) were assessed using transthoracic echocardiography one week after procedure, 6 moths and then annually after operation. These diameters were compared with normal valves values predicted by body surface area. V/s index (autograft annulus to sinus diameter) was assessed during follow-up. Z-score for autograft annulus was assessed just after operation(Z0) and in the late follow-up(Zf) and Z-score rate of change per year (g/y) was calculated.
Results: Z0 in group A was 3.02 ± 1.23 and in follow-up decreased to 2.3 ± 0.61(p = 0.01).
G/y index was -0.6 ± 0.9. In older patients in group B Z0 was 1.8 ± 0.74 and in follow-up increased to 2.9 ± 1.3 (p = 0.003). G/y index was 1.2 ± 1.7 (p = 0.001). In group A V/s index was 1.22 ± 0.12 and in follow-up didn't change significantly 1.25 ± 0.03.
In group B V/s index was 1.1 ± 0.07 and increased to 1.23 ± 0.05 in follow-up. There was no significant statistical difference in V/s index between 2 groups.
Conclusions: Autograft dilatation has been identified more often in patients who underwent Ross procedure above 7 yrs of age but it has not been associated with increased autograft valve insufficiency. Higher Z-score in younger patients was a result of physiological discrepancy in diameters between pulmonary and aortic valves.

O9-4
Perventricular, Non Surgical Aortic Reconstruction
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Objective: Treatment of aortic aneurysms is challenging in young adults with Marfan syndrome , often requiring surgical aortic reconstruction. Correction of the aortic valve along with support of the ascending and descending aorta might be necessary. Non surgical aortic reconstruction could conceivably offer palliation or repair, improving morbidity and mortality. The feasibility and safety of such a method was assesed in piglets.
Methods: Aortic reconstuction was performed in 8 piglets 10–14 kg. A perventricular needle puncture was performed, aiming at the loop of a pigtail catheter placed in the apex of the left ventricle. Polyurethane lined self-expandable stents (PLSES) were placed for the ascending aorta (5 cases, single stents), ascending and descending aorta (1 case with 2 stents), ascending aorta with incorporated aortic valve (1 case) and for the ascending aorta and transverse arch (1 case, one composite stent with multiperforated lining in the transverse arch). The aortic diameter was 12–14 mm and the stent diameter 15 mm. The stents were delivered through 14F sheaths introduced through the apex of the left ventricle. All procedures were performed under echocardiographic and fluoroscopic guidance; selective angiography and pressure recordings were obtained. The entry site was repaired by myocardial plugs and the animals were followed up to one month and had autopsies.
Results: Placement of PLSES was feasible in all attempted cases. All animals survived the initial procedure, except for one which developed ventricullar fibrillation after the composite ascending aorta and transverse arch stent obstructed the coronary arteries. One animal with ascending and descending aorta PLSES developed femoral artery obstruction. On long term all stents were patent without obstuction of the coronary arteries or neck vessels. Pathology demonstrated patent stents, free of thrombus with smooth lining (endothelialization). No significant pressure gradients were detected through the PLSES.
Conclusions: Non surgical perventicular aortic reconstruction is feasible, fast and relatively safe in piglets. Myocardial plugs are effective for a safe perventricular procedure.
Caution should be taken during placement to avoid obstuction to the coronaries, the aortic valve and the neck vessels. The method could have potential application in high risk for surgery patients.

O9-5
Twenty year review: Arterial Switch Operation cardiac angiography follow-up at OLHSC
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Introduction: The object of this study was to review patients who underwent cardiac angiography post arterial switch operation. Specifically our primary aim was to assess neo-aortic and neo-pulmonary valvar function, coronary anatomy and neo-aortic root dilatation. A secondary aim was to note the incidence of post-op complications amenable to intervention in the catheterisation laboratory.
Methods: We performed a retrospective chart review from 1985 –2005 of all patients who had undergone ASO.
Results: We identified 212 patients who had undergone ASO for TGA between 1985–2005. Medical record follow-up data was available on 184 of these patients.
Of the total number of medical records reviewed, 77.3% underwent follow-up angiography after ASO.
Pulmonary branch stenosis (PBS): 54% of patients at catheterization had PBS as determined on angiographic appearance as well as pressure gradient. (43.7% trivial-mild; 7.6% moderate; 3.4% severe).
Of the moderate to severe PBS (n = 13), further management included balloon angioplasty (n = 3), pulmonary branch stenosis stenting (n = 6) and surgical repair (n = 2).
PBS stenting was undertaken an average of 4.3 years from surgery with angioplasty an average 3.5 years post-op.
Aortic valve incompetence (AI): 26.9% of children at follow-up catheterization had trivial to mild AI; 4.2% moderate AI there was one case of severe AI. 3 out of the 5 cases of moderate AI had Taussig Bing anomaly.
All moderate AI on angiography was clinically detectable.
Neo-aortic dilatation: trivial to mild: 10.9%, moderate: 1.7% of caths. Half of cases with aortic root dilatation had concomitant AI (n = 8/15).
There was one case of severe fibromuscular sub-aortic stenosis.
Coronary angiography: Six patients had delayed or slow filling of their left coronary artery (5% of total). Five of these patients were symptomatic with symptoms of coronary insufficiency. The mean time post-operatively to angiographic diagnosis of poor LCA filling was 6 years.

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One patient underwent LIMA bypass grafting of her LCA stenosis.
Conclusions: Cardiac angiography remains a useful, safe and well-tolerated screening modality for long-term follow-up of patients after arterial switch operation.
Balloon angioplasty and stenting to developing pulmonary branch stenosis prevented or delayed a significant number of patients from further surgical management.

O9-6
ECMO-Transport: For the safe transport of critical ill pediatric patients
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Introduction: Critical ill pediatric patients often experience a further deterioration of their hemodynamic and pulmonary parameters during transport. Some patients are even too unstable for a transport with existing modalities so that they do not get the opportunity of maximal medical treatment. Our aim was it therefore to develop a ECMO-Transport-Program for the safe transport of theses critical ill patients.
Methods: We designed a small and lightweight ECMO stretcher for infants and adults, which fit in nearly every helicopter and in all ambulances. The Levitronix CentriMag centrifugal pump was chosen because of its lightweight consols and the long battery lasting. The ECMO circuit is safely secured on a rack at the end of the stretcher, so that patient and ECMO can be transported as one unit and the risk of disconnection is very low. We also build up and trained a small ECMO team, consisting of a pediatric heart surgeon, a pediatric cardiac intensivist, a pediatric cardiac intensive care nurse, a paramedic and a pilot.
Results: We started our ECMO-Transport-Program in March 2006 and until now we did seven ECMO-Transports. Three patients needed an emergency ECMO implantation by our team in other hospitals. The youngest patient needed emergency ECMO 3 hours after birth because of congenital diaphragmatic hernia. Another patient was a newborn baby with meconium aspiration syndrome in a small hospital in Austria, 600 km away from our ECMO centre. The third patient was a 5 years old boy with a univentricular heart palliated with a Fontan operation. He was resuscitated because of arrhythmia in a small hospital. After initial recovery he deteriorated again and venoarterial ECMO.
Three other ECMO-Transports were planned transfers of neonatal patients, which were already treated by ECMO and were transferred for surgical treatment to another ECMO centre.
All patients survived ECMO implantation and transport without incident. Five of seven patients were successfully weaned off ECMO.
Conclusion: The ECMO-Transports of our first seven patients were uneventful and the survival rate of ECMO was 71%. The small team and the lightweight technical equipment enable us to perform quick transports with helicopters or ambulances.

O10-1
Fetal echocardiography in the first trimester for fetuses at risk of congenital heart disease
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The purpose of this prospective study of the first trimester by echocardiography is to detect complex congenital heart disease (CHD) and to confirm the normality of the fetal heart. In this study 148 fetuses were examined for the first time at the mean age of 15 ± 1.5 weeks (12.2–17.6). All fetuses were controlled at least once after 18 weeks. We had 14 failures of examination, mean age 14.7 ± 1.6 weeks, mostly due to maternal overweight and anterior placenta. A total of 307 examinations were performed with a mean gestational age 17.5 ± 4.7 weeks (12.2–34.4). At the first examination patients were referred for nuchal translucency >3 mm (n = 112) with normal karyotype or for previous history of CHD (n = 36).
Results: We found 15 CHD in the nuchal group (11% of the population) and only one in the previous history group (n = 36) (recurence of hypoplastic LV). In the nuchal group, the CHD were conotruncal malformations (5, one with aortic hypoplasia), single ventricle (3, with pulmonary stenosis in 2), AVcanal (4, all umbalanced, with hypoplasia of the aorta in 2), fibroelastosis of the LV (1) and aortic atresia (2). We identified 2 fetuses who had a peculiar evolution: normal at 17 weeks, they evolved as hypoplasia of the aorta and small left ventricle at 26 and 28.6 weeks respectively. The statistics of the n = 148 young fetuses were: sensiblity of the method was 73%, the specificity was 98%, the predictive positive value was 85% and the predictive negative value was 97%.
Fetal echocardiography of the first trimester detects complex congenital heart disease early in pregnancy. However, care should be taken for the CHD flow dependent that can evolve in the second trimester. We recommend because of this possibility to perform a control in the second trimester whenever any discrete sign as asymetry appears at the control examination.

O10-2
Fetal left heart obstructions- diagnosis, development and outcome during the first year of life
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Introduction: Fetal heart disease may progress during intrauterine life. Our objective was to examine the cardiac characteristics, intrauterine development and first year outcome in fetuses with left heart obstructive lesions (LHO).
Objectives and Results: Therefore, we retrospectively evaluated complete datasets and video documentations of 55 fetuses which had been referred to our center between 1999 and 2006 because of suspected LHO. In 33 of them, hypoplastic left heart syndrome (HLHS) was diagnosed at a median gestational age of 22.5 weeks, in 12 of them critical aortic stenosis (AS, median age 25.5 weeks), and in 10 coarctation (CoA, median age 27 weeks).
Termination of pregnancy was performed in 7/33 cases with HLHS, intrauterine death occurred in one, and neonatal death due to denied surgery in seven. The remaining 18 patients underwent Norwood procedure with 3 deaths postoperatively due to restrictive foramen ovale or acute shunt thrombosis. Prenatal prediction of a restrictive foramen ovale was correct in 2 cases and false positive in 4.
Out of the 12 fetuses with AS, 10 had concommitant endocardial fibroelastosis. Three of them developed HLHS within 7 to 10 weeks and underwent Norwood operation postnatally; one of them died. In the remaining 9, ballon valvuloplasty was performed post partum. It resulted in severe aortic insufficiency in 2 and death in 1, and ended up in an univentricular approach in 3 of them (1 death).

Of the 10 patients with CoA, 6 had an hypoplastic aortic arch initially, 2 during later examinations. Surgery was necessary in 9/10, and all patients survived.
Conclusions: In this series, survival rate of fetuses with HLHS was 45.5%, of fetuses with AS 75%, and with CoA 100%. 25% of fetuses with AS ended up in HLHS before 30 weeks of gestation, and 25% in a Norwood procedure despite successful valvuloplasty postnatally. Thus, for optimal perinatal management, follow up intervals should be at least 4 weeks, and councelling should include developmental patterns.

O10-3
Results of stent implantation in 7 neonates with pulmonary atresia and left pulmonary artery stenosis via modified Blalock-Taussig shunts
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Objectives: The aim of the study is to assess the efficacy of stent implantation in palliative treatment of duct-dependent left pulmonary artery stenosis in neonates with pulmonary atresia.
Material and methods: Stents were implanted through modified right Blalock-Taussig shunts to relieve left pulmonary artery stenosis in 7 neonates: 1 pt with PA, 3 pts with PA+VSD, 1 pt with PA+VSD+TGA+LVOTO, 2 pts with PA and complex congenital heart disease. Mean age at the time of implantation was 23.7+10.9 days (range 12–42 days), mean body weight was 3342.9+799.7 g (range 1900–4200 g). Stents were implanted 7–31 days after shunt placement (mean 15.7+7.7 days). Palmaz Genesis premounted stents were used in 5 pts, premounted coronary stents in 2. Length of the stents was 12–15 mm. Mean stents diameter was 4.5+0.7 mm (range 3.5–5.3 mm).
Results: No procedure related complications were observed. One pt with pulmonary atresia and RV-dependent coronary circulation died 46 days after implantation due to LV dysfunction, 1 pt was lost from follow-up. Mean follow-up after the procedure of remaining 5 pts was 0.5–24+11.6 mths (range 0.5–24 mths), 4 of them have remained free from reintervention with laminar flow on colour-Doppler in the left pulmonary artery. One pt underwent successful stent redilation for neointimal proliferation 3.5 mths after implantation. 3 pts underwent surgical correction of the congenital heart defect at the age of 8–24 mths. In all cases stents were longitudinally cut and left in-situ in the left pulmonary artery and patch angioplasty of the artery was performed. One pt required stent implantation to both pulmonary arteries because of bilateral peripheral pulmonary arteries stenosis, and coil embolization of aorto-pulmonary collaterals in the early postoperative period.
Conclusions: Stents implantation through modified Blalock-Taussig shunts is a safe and effective palliative treatment of neonates with pulmonary atresia and duct-dependent left pulmonary artery stenosis. Second Blalock-Taussig shunt placement can be avoided in patients treated with this method.

O10-4
Catheterinterventional treatment of Sano shunt obstruction
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Introduction: Shunts placed between the right ventricle and the pulmonary arteries called Sano shunts recently modified Norwood surgery for hypoplastic left heart syndrome. Patients with Sano shunts tend to be more stable thus reducing the interstage mortality of this still challenging complex cardiac anomaly. However, Sano shunt stenosis may develop and is a life threatening complication. We report on our experience in patients with Sano shunt obstruction.
Methods: Retrospective observational report. Patients: Nine infants presenting with decreasing transcutaneous oxygen saturations (43–67%, median 60%) following modified Norwood procedures were shown to have relevant Sano shunt stenosis. None was suited for early stage-two surgery (cavopulmonary Glenn anastomosis). Catherization was performed at the age of 15 to 112 (median 71) days. Weight was 3.0 to 6.0 (median 4.7) kg. Technique: Femoral 5F venous access. The shunt was entered with a 4F right Judkins catheter and a selective angiography performed. The stenosis was localized proximal in 6, distal in 1 and proximal and distal in 2 patients. Eleven coronary stents were implanted.
Results: There were no procedure associated complications. Oxygen saturation increased immediately to 75–86% (median 80%) and remained above 70% during follow-up in all but one who stabilized only after a second stent was placed for additional intramural stenosis 37 days after the first stent implantation. Six patients had successful stage two surgery 48–288 (median 94) days after stent implantation. Catheterization prior to surgery showed no relevant in-stent stenosis. Surgery was not complicated by the stents. Three patients waiting for stage two surgery show stable saturations during close follow-up.
Conclusions: Sano shunt obstruction can be treated safely and effectively by stent implantation. In-stent restenosis seems not to be a problem in the limited time-span of Sano shunt dependence. Thus, early redo surgery can be avoided and stage-two operation can be scheduled electively.

O10-5
Aortic Dilation and Rearrangement of the Aortic Root in Turner Syndrome: Comparative Assessment Using Transthoracic Echocardiography and Magnetic Resonance Imaging
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Background: The prevalence of aortic root dilation in Turner syndrome (TS) patients with no predisposing cardiac lesion, and the rearrangements of the aortic root associated with aortic dilation are unknown.
Methods: We studied with transthoracic echocardiography and magnetic resonance imaging (MRI) 59 TS patients without associated cardiac lesions (median age 24 years, range 12 to 28 years) and 15 healthy volunteers. The diameter of the aorta at the level of the aortic annulus, sinuses of Valsalva, sino-tubular junction (STJ) and ascending aorta was measured by both imaging techniques. Aortic diameters were indexed for body surface area.
Results: TS patients had larger aortic annulus ($p = 0.004$), STJ ($p = 0.0001$), and ascending aorta diameters ($p = 0.01$) compared to control subjects. Dilation of the STJ was observed in 11/59 patients (18.6%) at echocardiography and in 8/59 patients (13.6%) at MRI. Dilation of the ascending aorta was observed in 9/59 patients (15.2%) at echocardiography and in 7/59 patients (11.9%) at MRI. Turner syndrome patients, especially those with a dilated ascending aorta, displayed a complex rearrangement of the aortic

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root characterized by prominent dilation of the STJ. An inverse association was noted between aortic diameters and age, and between STJ diameter at MRI and karyotype. Bland-Altman analysis showed a good agreement between measurements of the ascending aorta by echocardiography and MRI (bias –0.851 ± 2.11, 95% limit of agreement –4.99 to 3.30).
Conclusions: Dilation of the ascending aorta and of the STJ is relatively common in TS patients. Turner syndrome is associated with a morpho-metric reshaping of the aortic root with prominent dilation of the STJ. There is a good agreement between the diameters of the ascending aorta measured by echocardiography vs. MRI.

O10-6
Two years follow-up of the "growth stent" – results in infants with aortic coarctation
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Introduction: There is currently no stent available for possible curative transcatheter treatment of aortic coarctation in infants. To overcome this problem we designed a new stent, the "growth stent", which was tested to be not restrictive during growth in an animal experiment. We evaluated the feasibility and usefulness of the stent for the treatment of aortic coarctation in infants.
Methods: Prospective study of 13 growth stents implanted between April 2002 and January 2005. Analysis of immediate results and after a follow-up of 24 months (11 to 51 months).
Patients: Twelve patients aged 1–15 months (median 5 months). Body weight ranged from 3.4–12.8 kg (median 5.4 kg). Eight patients suffered from aortic (re-)coarctation, 4 of them from stenosis of the aortic anastomosis after a Norwood I procedure.
Results: Pressure gradients immediately after stent implantation dropped from 30 mmHg (range 20 to 50 mmHg) to 8 mmHg (range 0 to 15 mmHg). One patient died 11 months later of unrelated causes. Five patients had one (3pts) or two (2pts) balloondilations three to 28 months (median 12 months) after growth stent implantation. The median pressure gradient dropped from 25 mmHg (range 15 to 30 mmHg) to 15 mmHg (range 5 to 25 mmHg).
Six patients received a large stent after 19 to 34 months. Median body weight was 11.8 kg (9.4 to 15 kg) and implantation was performed through sheaths of 6F to 9F (median 8F).
Conclusions: The growth stent is suitable for the acute treatment of aortic coarctation in infants and can be overstented later on – whenever necessary – with a larger stent without causing restriction. Long term results with repeated redilations of overstented growth stents, however, are still lacking.

O11-1
From complications to changes in approach – experience with percutaneous pulmonary valve implantation
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Introduction: In our total experience of percutaneous pulmonary valve implantation (PPVI) we have encountered clinically relevant problems and complications during the procedure and follow-up period. Detailed analysis prompted changes in patient selection and alterations to implantation technique alongside changes in management strategies. We sought to report on this experience.
Methods: 137 patients (median age 18.0 years, range 7–71 years; 57 female) underwent PPVI between September 2000 and September 2006. Procedural and follow-up data were retrospectively reviewed.
Results: There was no procedural mortality. Immediate procedural complications developed in 6 patients (4%), all of whom required surgery, including valve-stent explantation in five and control of bleeding in one. 4/6 (67%) procedural complications could have been avoided in retrospect and led to changes in patient selection (valve dislodgement [n = 2], coronary compression [n = 1]) and implantation technique (RPA obstruction [n = 1]). However, homograft rupture [n = 2] remains impossible to predict. During follow-up (median 15.1 months, range 0.1–43.5 months) clinically relevant complications were identified in 27 patients (20%), the commonest of which was residual/re-stenosis in 25/27 patients, whereas 2/27 had endocarditis. Initially, all patients with re-stenosis underwent surgical valve-stent explantation (n = 14, including the two patients with endocarditis). Subsequently, repeat PPVI became the treatment of choice for specific indications ('Hammock effect' [n = 4], stent fracture [n = 8] and residual stenosis [n = 2]).
In another two patients PPVI was undertaken as a palliative treatment, since they presented 'in extremis'. Although both patients had uncomplicated procedures and good technical results, they died subsequently (one patient died 24 hours post-PPVI due to left heart failure and the other patient died two months post-PPVI due to overwhelming pneumonia). This represents the only mortality in our series. Whether application of PPVI in these conditions provides long-term clinical benefit as yet remains unclear.
Conclusion: Detailed scrutiny of the problems and complications encountered during implantation and follow up has allowed successful modifications to our approach.

O11-2
A new hybrid therapy for closure of muscular ventricular septal defects in a pig model
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Introduction: So far therapy for relevant muscular ventricular septal defects (mVSD) is challenging, especially in newborns and infants, because both – surgical and interventional therapies have their limitations. Hybrid therapy is a new innovative approach for mVSD's, which combines advantages of surgical and interventional techniques. We performed a pilot study to evaluate a novel technique for patch closure of mVSD in hybrid technique without extracorporeal circulation (ECC) in a pig animal model.
Methods: 5 pigs underwent anterolateral thoracotomy to expose the left ventricle. The mVSD's were created under 2D and 3D echocardiographic guidance (IE 33 Ultrasound system, Philips Medical Systems GmbH, Netherlands) with a 7.5 mm sharp punch instrument (n = 5), which was forwarded via a left ventricular incision to create a midmuscular or apical VSD, preferentially. A special designed patch system with a nitinol frame of 2 cm diameter (pfm, Cologne, Germany) was passed across the carotid artery over a 7 French long sheath into the left ventricle and positioned directly to the VSD. An instrument

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resembling a stapler for applying small nitinol anchors was introduced over a small ventriculotomy. The patch was fixed with anchors on the septum under echocardiographic and fluoroscopic guidance. Finally the nitinol frame was detached from the patch.
Results: The location of the defects was apical (n = 1), midmuscular (n = 3) and inlet muscular (n = 1). Closure of the mVSD was successful in four of five animals confirmed by echocardiography and hemodynamic measurements. Closure of the apical mVSD failed because the round shaped nitinol frame could not positioned directly to the apex. Dopamine (2–4 µg/kg/min) was administered when necessary. Animals were hemodynamic stable throughout the experiment.
Conclusion: The presented hybrid approach seems to be a feasible novel technique for closure of mVSD´s without using ECC. Further evaluation and development of the patch system is necessary to assess applicability for different conditions of mVSD´s in humans.

O11-3
Early experience with dilatable pulmonary artery band and interventional device closure of muscular ventricular septal defects
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Introduction: Banding of the pulmonary trunk continues to have a role in the initial management of some types of ventricular septal defect (VSD). We report early experience with the use of a dilatable pulmonary artery band used in patients thought to have clinically significant VSD's which, either might close spontaneously, or might subsequently be amenable to transcatheter closure.
Methods: Pulmonary artery banding was undertaken using 0.4 mm Gore-Tex tape secured by 2 surgical Ligaclips and sutured in position using 6/0 polypropylene to prevent migration. Previous in vitro experiments had shown this method of banding to be amenable to dilatation with a high pressure angioplasty balloon. Transcatheter 'debanding' was performed when echocardiograpy demonstrated that the VSD(s) had closed or when there was right to left flow across the VSD. It was combined with transcatheter VSD closure when necessary.
Results: 5 patients underwent pulmonary artery banding in the neonatal period, 4 of whom had simultaneous surgical repair of aortic coarctation. Balloon dilatation of the pulmonary artery band was performed at a median age of 8.4 months (range 6.0–10.9). 3 patients did not need VSD closure as the defects had either closed or were small; one of these had dilatation of recoarctation at the time of 'debanding'. One case underwent simultaneous transcatheter VSD closure and 'debanding'. One patient had partial dilatation of the band allowing more time for apical VSD's to become smaller and then underwent surgical debanding and VSD closure of a perimembranous defect too large for device closure. Transcatheter 'debanding' was undertaken using a high pressure angioplasty balloon (range 10–14 mm diameter) inflated with an indeflator. In all patients, transcatheter 'debanding' resulted in a marked reduction in the gradient across the band with the highest residual gradient of 30 mmHg in one patient who is awaiting further band dilatation.
Conclusions: Early experience using this technique would suggest that it is safe and effective for initial palliation in some types of VSD and that there is a predictable response to subsequent balloon dilatation of the band. It is recognised that further band dilatation will be required with growth which may be less predictable.

O11-4
Real-time Left Ventricular Physiology During Transcatheter Closure of Atrial Septal Defects Measured by Conductance Catheter
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Objectives: Transcatheter closure of an atrial septal defect (ASD) is a routine procedure today that even asymptomatic patients take advantage from. Rarely however, ASD closure is associated with acute lung edema and left ventricular (LV) dysfunction. It is unclear whether this is intrinsic and exposed by ASD closure, or whether it is caused by ASD closure.
Methods: 8 patients with ASD underwent transcatheter device closure of an ASD. Baseline LV systolic and diastolic functions were derived by analysis of endsystolic and enddiastolic pressure-volume relationships (ESPVR and EDPVR) both at rest and during preload reduction by transient balloon occlusion of the inferior vena cava, and during transient ASD closure with an ASD-sizing balloon. Analysis was carried out using a Millar® catheter and the CD Leycom® measurement kit and software.
Results: Pressure-volume loops showed a shift up and to the right immediately after ASD closure. Ejection fraction (EF) decreased slightly; however, ESPVR and EPPVR did not change.

	EDP (mmHg)	EDV (mmHg)	EF (%)	ESPVR	EDPVR
Baseline	7 ± 2.8	105 ± 23.5	59 ± 11.5	1.4 ± 0.4	0.3 ± 0.2
after ASD closure	11.5 ± 3.5	118.5 ± 19.6	55 ± 12.2	1.6 ± 0.6	0.2 ± 0.1
p	0.004	0.031	0.008	n.s.	n.s.

Conclusions: To the best of our knowledge, we showed for the first time the immediate adaptation of the LV during ASD closure in real time. The shift of the pressure-volume loops up and to the right immediately after ASD closure indicated an acute challenge in volume and pressure, accordingly. However, the finding that parameters of both diastolic and systolic LV function did not change would suggest unchanged intrinsic contractile myocardial function, indicating that interventional ASD-closure as such does not cause a change in myocardial function.

O11-5
Stent Implantation for the Treatment of Adult Aortic Coarctation: Initial and Five-Year Results
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Introduction: During the last decade there has been a significant number of reports of SI for the treatment of CoA. Although these studies report quite satisfactory angiographic and hemodynamic results they have a very short to intermediate-term follow-up. This study reports initial and 5-year results following stent implantation (SI) for adult aortic coarctation (CoA).
Methods: Thirty-one patients with CoA underwent SI (median age 35 years, range 23–58 years); twenty patients were treated for isolated CoA and eleven for recurrent CoA. Thirty-four stents were implanted. Palmaz 4014 stents were placed in

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22 patients, Palmaz 308 in eight. Elective re-dilation of previously implanted stent was performed in 6 patients with quite severe CoA.
Results: Immediately after SI the peak systolic gradient (mean (SD)) fell from 48 (18) mmHg to 5.3 (3.8) mmHg ($p < 0.05$). The diameter of the CoA increased from 5.8 (3.2) mm to 17.2 (3.8) mm ($p < 0.05$). The main procedural complication was proximal stent migration (catheter treated) in one patient. There were no early or late deaths, or any evidence of early or late aneurysm formation or any other complication related to SI throughout the follow-up period. At the 5-year follow-up no cases of recoarctation, were identified on angiography, multislice CT, or magnetic resonance imaging. In 84% (26/31) of the patients antihypertensive medication was either decreased or discontinued.
Conclusions: SI is an effective and safe alternative to conventional surgery for the treatment of adult patients with CoA.

O11-6
Stent implantation in aortic coarctation: Bare or Covered?
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Background: Bare stents have been used in all sites in patients with congenital heart diseases and large series and follow-up are reported in literature. However, even with these stents, aneurysms may form or aortic rupture may occur. Covered stents have been recently introduced for the treatment of aortic coarctation. We aimed to compare results and complications between bare stents and covered stents.
Methods and patients: Between January 2000 and December 2006, 104 consecutive patients (median age 15 years, range 6–66 years, 64 males) underwent cardiac catheterization for native or recurrent coarctation of the aorta. Seventy-two had native aortic coarctation, while 32 had recoarctation. Procedures were performed under general anhestesia. The following stents were used: Palmaz stents, Genesis stents, Cheatham-Platinum, covered Cheatham-Platinum. Bare stents were used in 71 patients, while covered stents were used in 33 subjects.
Results: There were no differences for age, gender, native coarctation/recoarctation rate, mean drop of peak systolic gradient, increase of diameter of coarcted segment, mean fluoroscopy and procedure times, between the two groups (BS: bare stents group vs CS: covered stents group).
Stents were placed in the correct position in all subjects in both groups. Long-sheath used for stent implantation was larger in CS group compared to BS group (median 12 vs 10 French; $p = 0.01$). Total complication rate was higher in BS (12% vs 0%; $p = 0.03$). The following complications occurred in BS group: early post-procedural death due to acute aortic dissection in 1 patient, stent embolization in 3 subjects, femoral artery pseudo-aneueysm needing vascular surgery in 3 pts, early periaortic hematoma in 1 pt, aneurysm formation during follow-up in 2 subjects.
Follow-up was longer for BS (median 38 vs 13 months; $p = 0.04$). A total of 6 subjects (4 in the BS group and 2 subjects in the CS group) needed re-dilation during follow-up. Finally, 10 subjects in BS (14%) and 9 in CS (27%) needed anti-hypertensive drugs during follow-up ($p = 0.14$).
Conclusions: Covered Cheatham-Platinum stents are promising tools for the treatment of aortic coarctation.
They appears to be safer that bare metal stents. Long-term follow-up data are needed.

O12-1
Presence of accessory pathways in the developing human heart, possible explanation for fetal and neonatal atrioventricular reentrant tachycardias
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Introduction: Fetal and neonatal atrioventricular reentrant tachycardias can be life threatening but resolve in the majority of cases during the first year of life. The temporary presence of accessory atrioventricular (AV) myocardial pathways during the normal process of isolation of the AV junction may explain this phenomenon.
Methods: We studied 44 human embryonic ($n = 6$), fetal ($n = 34$) and neonatal ($n = 4$) sectioned hearts with an age range of 4 to 36 weeks of development. (Immuno-)histochemical markers were used to identify myocardium and connective tissue, including antibodies against MLC-2a, HHF-35, collagenVI and periostin. Accessory AV myocardial pathways were quantified and categorized according to their specific location and 3-D AMIRA reconstructions were made.
Results: Up to 6 weeks of development, the atrial and ventricular myocardium was continuous at the AV junction. Between 6 and 10 weeks, numerous accessory myocardial pathways were observed at the left (45%), right (35%) and septal (20%) region of the AV junction. Whereas the most right sided accessory AV connections were identified as distinct myocardial strands, the left sided AV connections comprised larger areas of myocardium. Between 10 and 20 weeks, the accessory AV connections all consisted of discrete myocardial strands and gradually decreased in number. The majority of accessory connections (67%) were observed at the right AV junction, mostly located at the right lateral aspect of the right AV junction (45%) in close contact with the so-called right atrioventricular ring bundle. At the left AV junction and the septal area only 17% and 16% of the accessory AV connections were observed, respectively. 3-D reconstructions of the AV nodal area at these stages also demonstrated multiple accessory AV connections related to the developing AV node. From 20 weeks until birth and in neonatal hearts no more accessory AV connections were observed.
Conclusions: The isolation of the AV junction is a gradual and ongoing process and particularly right lateral pathways are commonly found at later stages of normal human cardiac development. These transitory accessory pathways may act as substrate for atrioventricular reentrant tachycardias in the fetus or neonate.

O12-2
Impact of the matrix used in tissue-engineered heart valves on bacterial adhesion in a model of endocarditis
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Patients requiring heart valve prosthesis are at high risk for acquiring endocarditis. Currently new heart valves based on the principle of tissue-engineering are under construction to mini-

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mize the risk factors and improve the biocompatibility. A tissue-engineered heart valve consists of a basic matrix seeded with autologous endothelial cells. Different materials are under investigation to serve as a matrix, particularly fibrin gel.
The impact of the different matrices on the probability of bacteria to infect endothelial cells which are seeded on these matrices is not known yet. In the present study we investigated the influence of fibrin gel matrix versus polysterene plates on the degree of bacterial adhesion and invasion which is an initial step of the bacterial-endothelial interaction and essential for the infection process.
Staphylococcus aureus, Streptococcus sanguis and Staphylococcus epidermidis, three relevant bacteria causing endocarditis in children, were used in the infection experiments.
Human venous endothelial cells were cultivated on fibrin gel and alternatively on polystyrene tissue culture plates. After grown to a confluent monolayer cells were incubated with S. aureus, S. sanguis and S. epidermidis.
Endothelial cells incubated with S. aureus showed a higher infection rate cultured on fibrin gel ($4.0 \pm 0.91\%$ of the inoculation dose) than those cultured on tissue culture plates ($1.44 \pm 0.42\%$). A higher infection rate was also found for endothelial cells incubated with S. sanguis and S. epidermidis being cultured on fibrin gel vs. tissue culture plates ($0.56 \pm 0.18\%$ vs $0.17 \pm 0.05\%$ and $3.3 \pm 1.36\%$ vs $1.3 \pm 0.48\%$ of the inoculation dose). There was a significant difference between the infection rates for S. aureus and S. sanguis ($p < 0.01$ and $p < 0.05$).
We conclude that the matrix serving as a basic structure for tissue-engineered heart valves has an impact on the risk of endocarditis. Recent studies emphasize the significant contribution of bacterial adhesion in inducing proinflammatory endothelial responses. Therefore it could be suggested that the matrix has also an influence on endothelial activation. Further investigations might state the importance of different matrices as inducers of pathways to evoke inflammation, tissue damage and fibrin deposition at the infected endovascular sites.

O12-3
Differential proteomic analysis of the pressure-overloaded right ventricle of young rats over time reveals specific changes in heat shock- and antioxidant proteins

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Introduction: In many forms of congenital heart disease, the right ventricle (RV) is subject to abnormal loading conditions resulting in RV hypertrophy and remodelling. We determined the alterations in RV proteomic phenotype in rats after prolonged RV pressure overload.
Methods: Male Wistar rats (8wk) underwent PAB or sham operation and were studied 6, 12 or 20 wks later ($n \geq 6$ per group). Parameters of RV contractility were assessed with biventricular pressure-volume loops. For proteomics, individual RV homogenates were subfractionated and the cytoplasmic fraction was subjected to 2-DE (pI 3–10, Mw 10–250 kDa). Coomassie blue stained gels of individual animals were compared using PDQUEST. Significantly up- or downregulated spots were in-gel trypsinized and analysed by MALDI/TOF-MS linked to Mascot.
Results: In PAB, RV systolic pressure was increased to 60% of systemic systolic pressure ($p < 0.05$), resulting in RV hypertrophy (RV/BW ratio 1.78, 1.80 and 1.83 fold increased resp. as compared to sham, $p < 0.05$). RV load-independent contractile parameters (Ees, PRSW, dP/dt max-Ved) were increased in PAB ($p < 0.05$) and were compatible with compensated RV hypertrophy in all three time groups. Proteomic analysis identified a group of 72 spots that differed in intensity between sham and PAB in at least one time group. Overall, the majority of these RV protein changes were metabolically related indicating a shift towards the glycolytic pathway at the expense of theß-oxidation. Amongst others, we identified a change in the heterodimeric composition of enolase over time. In addition, numerous stress-related proteins were altered in the RV of PAB. These included a time-dependent increase in expression of several (phosphorylated) HSP-27 species in PAB ($p < 0.05$). The increase in phosphorylated HSP-27 could not be explained by activation of the p38-MAPK pathway. In addition, alterations in the antioxidant proteins peroxiredoxin type 2 and 6 were observed.
Conclusion: Our data show that during the time course of compensated RV hypertrophy there is activation of several components of the protein quality control machinery, including the defence against oxidative stress. These protein changes are likely to be a part of a protective mechanism against the development of RV failure.

O12-4
Stem Cells recruitment in Surgical palliation for Hypoplastic Left Heart Syndrome

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Introduction: The recognition that myocytes and vascular structures could be formed from primitive cells homed to myocardium from circulation or from resident cardiac stem cells was accompanied by the idea that increased cardiac mass in cardiac hypertrophy could result from a combination of myocyte hypertrophy and hyperplasia. Hypoplastic Left Heart Syndrome is a complex congenital heart disease, characterized by severe hypoplasia of the left-sided heart and following normal surgical palliative procedures. Aim of the study was to identify the role of cardiac stem cells in the right hypertrophy remodelling in a group of patients affected by Hypoplastic Left Heart Syndrome.
Methods: Eleven Hearts of the affected patients who underwent surgical palliation procedures (with ages ranging form 9 to 365 days) were compared with age-matched controls. Samples taken from the inflow and outflow tratcs of the right ventricle were evaluated at histology and immunohistochemistry for stem cells markers (i.e. CD117, CD105, MDR1).
Results: we observed a recruitment of cardiac precursor stem cells CD117 and CD105 positive, both in the inflow and the outflow tract in operated hearts compared to the controls (CD117 inflow: controls 7.8 ± 3.3, operated hearts 18.09 ± 15.85 cells/field, with $p = 0.059$; outflow: controls 9 ± 4.73, operated patients 17.81 ± 13.61 cells/field with $p = 0.01$; CD105 inflow: controls 1.3 ± 0.9, operated patients 9.18 ± 3.8 cells/field, $p = 0.07$; outflow: controls 1.8 ± 1.6, operated patients 6.2 ± 3.1 cells/field, $p = 0.1$). Moreover cardiac precursor stem cells seem to be mainly mobilized by acute injury (early death patients) rather than by chronic processes (late death patients) since a higher presence of putative cardiac stem

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cells (CD105) was detected (acute 10.75 chronic 4.1 cells/field) in operated patients. Putative cardiac stem cells are mainly located in the interstitium and near collagen III struts. Rare cycling cardiac precursor cells, positive both for myosin and Ki67 markers were found.
Conclusions: During right ventricle hypertrophy we observe a recruitment of cardiac stem cells, in particular of endothelial precursor progenitor cells in cardiac remodelling. The latter the precursor stem cells could be primarily involved in the interstitial and endothelial tissues remodelling.

O12-5
Extracardiac Progenitor Cells Repopulation in Pediatric sex mismatch heart transplants
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Introduction: In humans, the differentiation of progenitor cells into cardiomyocytes has been studied in allografted female hearts transplanted into male recipients by examining the presence of Y-chromosome-positive cells. The data suggest that adult progenitor cells of non-cardiac origin may translocate from the recipient to the graft. It is plausible, however, that some of the male cells identified within a female heart may be of fetal origin, resulting in a microchimerism.
Aim of study was to evaluate the chimerism phenomenon of mismatched heart transplants in pediatric patients in order to assess the role of pregnancy microchimerism events.
Methods: Endomyocardial biopsies (n = 10) were obtained from the right ventricles of pediatric male patients (n = 5, mean age 15.2 ± 3.01 years) who had undergone sex-mismatched orthotopic heart transplantation.Cells from recipient origins were identified by FISH with a combined XY-chromosome probe. Biopsies at 2 different time points were examined in each patient. Rejection scores (according to ISHLT grading system) were calculated for all patients.
Results: Cardiomyocytes of recipient origin were detected in the major of biopsies with a mean percentage of 0.35 ± 0.15%. Cardiomyocytes of male origin (myosin positive, and CD45 negative) were detected in normal myocardium tissue as single cells (and not found in clusters).The nuclei of these Y-chromosome positive cardiomyocytes never showed more than two signals, one for Y-chromosome and one for X-chromosome. Multivariate statistical analysis showed an increase of recipient-derived cells in the donor hearts over time ($p < 0.05$).
Conclusions: Our results confirmed the presence of Y-chromosome cardiomyocytes in pediatric patients, excluding any maternal microchimerism influence, and with a phenotic transformation due to transdifferentiation and not to cell fusion.

O12-6
Exercise training does not improve significantly endothelial function in pre-pubertal obese children but prevent intima media thickening
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Background: We have previously described in pre-pubertal young obese children, compared with lean subjects, impairment of endothelial function without modification of mechanical properties and no increase of intima media thickness (IMT).
Aim: The present study aimed to assess the effect of an exercise training program on vascular function in a sub-group of these patients.
Study design: Forty three pre-pubertal obese subjects (age = 8.7 ± 0.2 year-old) were assigned to an exercise (ET) or non-exercise (NET) protocol in a randomized way. Twenty two (age = 8.6 ± 1.7 year-old, body mass index (BMI) 25 ± 5 kg/m^2) were not submitted to exercise training, while the influence of 12 weeks of moderate exercise training (3 sessions per week, 60 minutes per session) was examined in 21 obese subjects (age = 8.9 ± 1.4 year-old, BMI 25 ± 4 kg/m^2). We assessed the effect of exercise on IMT, mechanical properties of the CCA and flow-mediated dilation of the brachial artery (FMD), an endothelial dependent function, using high-resolution ultrasound.
Results: ET group obese children showed a slight decrease of BMI (24.3 ± 3.5 kg/m^2). Mechanical properties of the CCA remained not altered. FMD increased not significantly (6.0 ± 2.9% vs 5.8 ± 2.2%, P = 0.8). NET group obese children showed a non significant increase of body weight and BMI. IMT increased significantly from the baseline value (0.48 ± 0.3 mm vs 0.50 ± 0.03 mm, P < 0.02) without concomitant modification of mechanical CCA parameters and FMD.
Conclusion: Obese pre-pubertal children submitted to an exercise program showed a trend to improve brachial artery FMD, a validated surrogate measure of early atherosclerosis. Absence of exercise influences IM thickening.This study supports the value of an exercise program, as a secondary prevention setting, to reduce the progression of obesity and its vascular consequences.
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O13-1
Prognostic Value of B-Type Natriuretic Peptide in Children with Pulmonary Hypertension
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Background: B-type natriuretic peptide (BNP) is an established marker of myocardial dysfunction and has prognostic implications in different cardiovascular cohorts. We aimed to assess whether BNP relates to functional status and outcome in children with pulmonary hypertension (PH).
Methods and Results: BNP was measured in 50 children (male: female, 1.8:1) with PH aged 8.4 ± 5.1 years (range 0.3–18.4). Twenty-seven patients were diagnosed with idiopathic pulmonary arterial hypertension (IPAH), while 23 patients had associated PH [congenital heart disease (n = 17), lung disease (n = 4), other (n = 2)]. Functional clinical status (WHO-Functional Class), six-minute-walk test, echocardiographic and haemodynamic data were assessed. Mean BNP value was 144 ± 236 pg/ml (range <5–1250). There was no significant difference between BNP values in patients with IPAH or associated PH (p = 0.36). BNP correlated with WHO Functional Class (38 ± 53; 222 ± 314; 280 ± 277 in class II, III, and IV, respectively; p = 0.03), with echocardiographic assessment of right ventricular function (p < 0.0001), hypertrophy (p < 0.001) and dilatation (p < 0.0001), and in IPAH to haemodynamic status (mean pulmonary arterial pressure [PAP] at baseline and PAP and the pulmonary vascular resistance index [PVRI]

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with 60% FiO2 and 20ppm inhaled nitric oxide; $p < 0.05$ each). During a mean follow-up of 14.0 ± 7.5 months (range 0.2–33.1) 7 patients died, 5 children underwent transplantation and 2 were listed for transplantation. A BNP value >130 pg/ml was identified as the optimal cut-off value predicting a combined end-point of death, transplantation or admission to the active transplant list ($p < 0.04$). However, six children who died or were transplanted had a BNP lower than this value. This reflects the limited sensitivity of BNP (57.1%) for predicting death or need for transplantation in children with PH.

Conclusion: BNP correlated with functional status, echocardiographic and haemodynamic measures and outcome in a cohort of children with PH. BNP may be a useful adjunct to the assessment of clinical status but should not be used in isolation as independent screening tool to predict outcome in this cohort.

O13-2
Beneficial effects of ghrelin in a new model of infantile pulmonary hypertension induced by monocrotaline

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Introduction: Pulmonary hypertension (PH) is characterized by structural changes of the pulmonary vessels as well as right ventricular (RV) hypertrophy. The reversibility of pulmonary vascular remodelling is possible only at paediatric ages. The characterization of the infantile PH model will allow the study of pathophysiology of this disease and to test the effects of new drugs. Ghrelin (GHR) is a new vasoactive peptide with several cardiovascular effects. The aim of the study is to establish an infantile MCT-induced PH model and to evaluate hemodynamic and morphometric effects of administration of GHR.

Methods: Wistar rats 8-day-old randomly received MCT (30 mg/kg, sc) or equal volume of vehicle (Group Sham, n = 10). At D7, animals treated with MCT randomly received GHR (100 g/kg, sc, bid) (Group MCT-GHR, n = 10) or saline (Group MCT, n = 8) for 2 weeks. At D21, animals were instrumented to record several biventricular hemodynamic parameters. At the end, heart and lungs were excised and weighted.

Results: Presented in table as mean ± SEM. $P < 0.05$: a vs Sham; b vs MCT. Survival studies at D21: Sham = 100%, MCT = 85%, MCT+GHR = 100%. No differences were found in LV hemodynamic parameters.

Conclusions: We characterized, for the first time, the biventricular hemodynamics of healthy Wistar rats during the childhood period and established a model for paediatric MCT-induced PH. Animals treated with MCT presented RV hypertrophy, PH and diastolic dysfunction (elevation of end-diastolic pressure; slower myocardial relaxation rate) in RV. Ghrelin administration increased survival rate and ameliorated PH and RV hemodynamics. These beneficial effects of GHR open new therapeutic options for this disease in paediatric population.

	Sham	MCT	MCT-GHR
Body Weight (BW), g	80 ± 1	58 ± 3a	108 ± 3a,b
Lung/BW, g/kg	8.20 ± 0.33	11.534 ± 0.82a	8.77 ± 0.30b
RV/BW g/kg	0.741 ± 0.07	1.735 ± 0.16a	1.414 ± 0.12a,b
LV/BW g/kg	3.27 ± 0.45	4.36 ± 0.26 a	3.15 ± 0.11b
RVPmax, mmHg	19.6 ± 2.8	37.1 ± 4.7a	22.6 ± 3.2b
dP/dtmax RV, mmHg/s	663 ± 74	1210 ± 258a	908 ± 187
RVEDP, mmHg	0.38 ± 0.7	2.9 ± 1.4a	1.81 ± 0.43b
Tau, ms	13.2 ± 0.8	28.3 ± 4.3a	12.8 ± 1.1b

RVPmax – peak systolic pressure; RVEDP – end-diastolic pressure; dP/dTmax – peak rates of ventricular pressure; Tau – myocardial relaxation rate.

O13-3
Parameters of heart rate variability predict outcome in children with pulmonary hypertension

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Background: Prognosis for children with pulmonary hypertension (PH) remains poor, and sudden death is common. Objective risk stratifiers are required to assess need escalation of medical therapy or transplantation. Autonomic dysfunction is associated with cardiac death in various cardiovascular cohorts. We hypothesized that heart rate variability (HRV) – as an established measure of cardiac autonomic nervous function – may also predict outcome in children with severe PH.

Methods: Forty-seven consecutive patients with severe PHn (27 male; mean age 11.2 ± 5.4 years) were included in this study. Twenty-four patients were diagnosed with idiopathic pulmonary arterial hypertension, while 23 patients had associated PH.

The following parameters of HRV were determined from Holter electrocardiograms: standard deviation of normal-to-normal intervals (SDNN), standard deviation of mean values for normal-to-normal intervals over 5 minutes (SDANN), and square root of the mean square differences of successive RR intervals (RMSSD).

In addition, functional clinical status (WHO-Functional Class), six-minute-walk test distance, echocardiographic and haemodynamic data were assessed.

Results: Children who died or underwent transplantation had significantly lower values of SDANN (70.7 ± 30.9 vs. 119.2 ± 57.2, $p = 0.006$), SDNN (79.5 ± 30.5 vs. 129.9 ± 63.6, $p = 0.009$), and RMSSD (22.3 ± 11.7 vs. 47.7 ± 34.5, $p = 0.015$) compared to the remaining patients. During a mean follow-up period of 16.8 months (range 1.8–45.6 months) 8 patients died and 5 underwent transplantation. On univariate Cox proportional-hazards analysis all parameters of HRV predicted death or need for transplantation ($p < 0.05$ for each). In addition SDANN and SDNN were also significantly predictive of mortality alone ($p < 0.05$ for each). On multivariate analysis SDANN and SDNN predicted outcome independently of functional status, history of syncope, right ventricular function, and haemodynamic parameters such as pulmonary vascular resistance with and without nitric oxide. Patients with a SDANN-value below 100 had a 4.5 fold increased risk of death or need for transplantation in our study (event free survival at 24 months: 90 vs 40%; $p = 0.03$).

Conclusions: Holter monitoring is widely available and is a noninvasive cost-effective tool to measure HRV. Our study demonstrates for the first time that parameters of HRV predict

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survival in children with PH. Heart rate variability may represent a promising adjunct to assess prognosis in this population.

O13-4
The use of Bosentan in the treatment of Pulmonary Arterial Hypertension in infants less then one year of age

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Introduction: Bosentan, a dual endothelin receptor antagonist, has been shown to be an effective agent for the treatment of pulmonary arterial hypertension (PAH) in children and adults. We report on the use of Bosentan in infants less than 1 year of age in our institution between 2003 and 2006.
Methods: We performed a retrospective chart review in all infants treated with Bosentan, specifically focusing on efficacy, safety and tolerability.
Bosentan was prescribed for pulmonary hypertension confirmed by echocardiography or cardiac catheterisation in patients with either PAH as a pre-operative contra-indication to surgery or symptomatic disease.
All survivors were followed up at our institution with serial liver function test screening, out-patient assessment and follow-up echocardiography.
Ethics committee approval was obtained for this study.
Results: Between 2003–2006, 18 infants received Bosentan for documented pulmonary arterial hypertension. Medical records were incomplete on one infant.
The primary pathology was congenital heart disease (n = 14), parenchymal lung disease (n = 2), and idiopathic pulmonary arterial hypertension (n = 2).
The average dose of Bosentan commenced was 1mg/kg b.d. at an average of 4.9 months of life. The average duration of therapy was 6.3 months.
In addition to Bosentan anti-hypertensive therapy, 94% of the infants also received sildenafil, 29% phenoxybenzamine, 23% epoprostenol and 47% inhaled nitric oxide with a mean peak dose of 27.8ppm.
3/18 (16.6%) of infants had a mild rise in serum transaminases (defined as less than 250U/L). One critically ill infant developed marked liver impairment during the course of ICU admission (AST 2401, ALT 895) prompting cessation of Bosentan. Transaminases returned to normal range on discontinuation of therapy.
12 infants (66.6%) survived to discharge. 10 infants were cared for in ICU with an average length of ICU stay in the survivors of 25.8 days.
7 infants commenced Bosentan as out-patients.
At last follow-up, average weight of (non-syndromic) survivors was on the 30th centile with an average NYHA class PAH score of 2.2. Home oxygen was being used by 3 infants.
Conclusions: Bosentan is a generally well tolerated and safe drug for the treatment of pulmonary hypertension of varying aetiologies in infants under one year of age.

O13-5
Cardiac Autonomic Nervous Activity is Impaired in Patients with the Eisenmenger Syndrome and Tends to Improve Under Treatment with Bosentan

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Introduction: While it has been shown that heart rate variability (HRV) is reduced in PAH and Fallot's tetralogy, this has not been studied in Eisenmenger syndrome (ES) yet. Furthermore, we wondered whether these indices change in response to pulmonary vasodilator therapy with the endothelin antagonist bosentan (Tracleer®).
Methods: Before and after 26 weeks of therapy, continuous ECG and simultaneous non-invasive beat-to-beat finger cuff blood pressure measurements were taken during 10 minutes of spontaneous breathing and with a set rate of 10s/breath for another 10 minutes. Spectral domain analysis of heart rate and blood pressure variation were performed using the CN Systems® software. Additionally, all subjects underwent 24h-Holter recordings for time domain analysis.
Results: 11 adult patients (5 male, mean age 31.2 years) with ES, all in sinus rhythm, were examined.

Variable	LF/nu	HF/nu	LF/ms2	HF/ms2	Tot pow ms2
ES; base	42 ± 16	58 ± 16	140 ± 135	232 ± 246	503 ± 437
ES; treat	42 ± 19	58 ± 19	145 ± 110	355 ± 482	612 ± 585
p-value: Rx	0.85	0.84	0.93	0.46	0.49
normals	54 ± 4	29 ± 3	1170 ± 416	975 ± 203	3466 ± 1018
p-value: ES	0.04	0.01	<.01	<.01	<.01

Variable	LF dBP/HF	SDNN	SD-ANN	TI	rmsSD	pNN50
ES; base	0.7 ± 0.3	125 ± 53	109 ± 55	21 ± 9	66 ± 22	11 ± 9
ES; treat	0.8 ± 0.5	145 ± 50	118 ± 40	26 ± 15	111 ± 35	19 ± 14
p-value: Rx	0.53	0.06	0.54	0.11	0.08	0.06
normals	<2.0	152 ± 52	112 ± 42	37 ± 15	27 ± 12	26.9 ± 22.8
p-value: ES	<.01	ns	ns	0.04	ns	0.02

Conclusions: Here we show for the first time that HRV is severely impaired in ES patients. There was a trend to increasing HRV after 26 weeks of bosentan treatment encouraging for a further measurement after one full year. Further studies to correlate these findings with clinical parameters are under way.

O13-6
Early Predictors of Cardiomyopathy in Adolescents with Type I Diabetes

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Background: Diabetic cardiomyopathy has been suggested as a cause of cardiovascular morbidity in diabetics.
Objective: We aimed to assess adolescents with type I diabetes for early echocardiographic signs of subclinical cardiac dysfunction and to assess the value of N Terminal pro-Brain natriuretic peptide (NT- pro BNP) in detecting such cases.
Methods: 60 adolescents were studied; 40 diabetic patients (mean age 14.9 ± 2.04 years) and 20 healthy age and sex matched controls (mean age 14.3 ± 1.4 years) with normal clinical examination and no history of cardiac disease. All were assessed by echocardiography for chamber dimensions, systolic and diastolic function including myocardial performance index (MPI) and tissue Doppler interrogation of cardiac walls. Serum level of NT-pro BNP was measured.
Results: All diabetics and controls had normal cardiac dimensions and systolic functions (ejection fraction EF% 64.6 ± 8.4 versus

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65 ± 7.8 p > 0.05, shortening fraction SF% 35.3 ± 6.7 versus 35.9 ± 5.8 p > 0.05). Diastolic dysfunction was detected in 10 (25%) diabetics by conventional Doppler with a higher peak A 0.57 ± 0.15 versus 0.48 ± 0.09 (p < 0.05) and a lower E/A 1.6 ± 0.45 versus 2.09 ± 0.41 (p < 0.05) compared to controls. Diabetics showed a longer isovolumic contraction time (IVCT) 64.4 ± 17.6 versus 57.3 ± 8.3 (p < 0.05) and MPI 0.57 ± 0.15 versus 0.48 ± 0.10 (p < 0.05), in contrast to a shorter ejection time ET 242.7 ± 34 versus 263 ± 19 (p < 0.05) compared to controls. Isovolumic relaxation time IVRT was longer in diabetics than controls 74.34 ± 22.8 versus 64.5 ± 21.5 with no statistical significance. Tissue Doppler showed delayed myocardial relaxation in 21 (52.5%) diabetics with lower peak E 0.12 ± 0.03 versus 0.15 ± 0.03 (p < 0.01) and E/A 2 ± 0.8 versus 2.6 ± 0.6 (p < 0.01) compared to controls. NT-pro BNP was higher in diabetics (mean 65.5 ± 18.3) compared to controls (p < 0.01). NT-pro BNP levels correlated inversely with Peak E (r −0.32; p < 0.05) and E/A (r −0.42; p < 0.05) and positively with peak A (r 0.47; p < 0.05) with a cut off value of 62.5, 82% sensitivity, 95% specificity and 91% predictability.

Conclusion: Young diabetics already have significant delayed myocardial relaxation. Tissue Doppler has an additional value in the evaluation of ventricular filling in such patients. NT-pro BNP may be considered as a sensitive predictor, however further studies on larger scales are recommended.

O14-1
Transcatheter ASD closure in children below 13 kg

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Introduction: Transcatheter closure of secundum type atrial septal defect (ASD) has become a treatment of choice in selected patients. Such therapy in smaller children is still controversial but is indicated occasionally in special clinical circumstances. Experience of two tertiary centers is presented

Material and methods: Among 812 procedures of transcatheter closure of ASD, there were 67 children aged from 0.2 to 4.7 years, with body weight between 3.1 and 13 kg. These patients needed earlier treatment because of failure to thrive and/or recurrent respiratory tract infection and/or heart failure. In 3 children, there was significant residual shunt through ASD after previous complex surgery, 2 other children with PA with IVS had a right to left shunt through an ASD after previous neonatal Brock and Blalock Taussig operations. Mean ASD diameter (TEE) in the whole group was 8.3 (4–12), stretched diameter was 13 (5–18) mm. In 25 children, balloon sizing of ASD was omitted. There were 66 Amplatzer devices (ASO) deployed with size ranging from 5 to 18 mm and 1 Starflex umbrella of 23 mm in case of multiperforated aneurysm of IAS.

Results: The procedures of transcatheter ASD closure with ASO were successful in all but one patients, without residual shunt. There were no complications observed during and after the procedures. During mean follow-up of 3.3 years, range 0.1 to 9.2 years, clinical improvement was observed in all patients. The cause of unsuccessful ASO implantation in the smallest child of 3.5 kg was oblique position of ASO inside the defect despite many attempts at repositioning. Fluoroscopy time ranged from 0.5 to 40 (mean 10) min. In both pts with R-L shunt through ASD, cyanosis disappeared after the procedure with a permanent increase of saturation to 96%.

Conclusion: Transcatheter ASD closure in children <13 kg of body weight is an effective although technically more demanding mode of treatment.

O14-2
Results of transcatheter closure of unroofed coronary sinus

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Introduction: An unroofed coronary sinus allows shunting of blood flow between left atrium and coronary sinus. We present the results of transcatheter closure of the defect with Amplatzer septal occluder.

Methods: During a 2.5-year period, a total of 6 patients (4 males and 2 females) with ages ranging from 26 to 56 years underwent attempted transcatheter closure. All 6 patients were symptomatic. Prior to the procedure, a computerized tomography was performed to delineate location and size of the defect and presence of persistence of left superior vena cava. Transcatheter closure of unroofed coronary sinus was performed under general anesthesia and transesophageal echocardiographic (TEE) guidance.

Results: No patient had associated persistent left superior vena cava. The mean Qp/Qs ratio was 2.5 ± 1.1. Pulmonary hypertension was present in 2 patients. The maximal defect diameters on TEE images ranged from 13.5 to 20 mm. (mean 16.3 ± 2.5 mm) Amplatzer septal occluder was deployed in all 6 patients. The device was deployed at the ostium of coronary sinus in 5 and in the defect in remaining 1 patient. The mean diameter of device used was 20.3 ± 2.3 mm. (ranging from 16 to 22 mm) No residual shunt was found in any patient in the most recent echocardiography. After a mean follow-up period of 12.8 ± 6.9 months, no patient developed arrhythmia. All 6 patients had improvement in symptoms.

Conclusions: In the absence of persistent left superior vena cava, transcatheter closure of unroofed coronary sinus using Amplatzer septal occluder is feasible.

O14-3
Effectiveness of transcatheter patch release with surgical adhesives in the occlusion of heart defects

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Objective: Surgical adhesives containing polyethylene glycol (PEG) have been found experimentally to accelerate transcatheter patch release (TP) compared to natural fibrin in the occlusion of heart defects. Purpose of this study was to assess the effectiveness and safety of accelerated patch release using PEGs in a variety of heart defects in humans.

Methods: Twenty five heart defects or cardiac structures in patients 1–60 years old were treated using surgical adhesive attacchment of the transcatheter patch. Heart defects included 10 atrial septal defects (ASDs) 15–27 mm in occluding baloon diameter, 5 membranous ventricular septal defects (VSDs) 5–11 mm in angiographic diameter, 5 patent ductus arteriosus (PDA) 8–15 mm in angiographic diameter and 5 cases of left atrial appendage (LAA) 18–24 mm in echocardiographic diameter at the mouth of the LAA. The surgical adhesive was applied as a

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thin film of inactive solution (solution A-acid) for each particular application. Activation was delayed to allow catheter manipulation and patch placement with later adhesive activation (solution B-alcali or exposure to blood). A 50% test balloon deflation before final release was performed. The immediate results were assessed by fluoroscopy, trans-esophageal echocardiography and/or cine-angiograpy. Follow up was reported.
Results: TP with PEG adhesive film was succesful in all heart defects with the exception of 2 VSDs. The TP was well attached in all defects up to 25 mm in diameter. In one 27 mm ASD, TP was not well attached and was extracted percutaneously. Another 27 mm ASD with a residual shunt on 50% was left for an additional 8 hours prior to release with good result. Two VSDs had problems in advancing and releasing the patch due to tortuous and kinked introducing sheath, resulting in significant residual shunt in one case. No serious complications were reported. On follow-up all patients are doing well with the exception of the VSD case which required surgical correction for the residual shunt.
Conclusions: TP placement using surgical adhesives (PEGs) is effective and safe for the majority of heart defects up to 25 mm.

O14-4
Use Of Stents In Patients With Congenital Heart Defects
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Material and methods: 176 stents were used in 169 patients with different congenital heart diseases. To treat 85 stenosed segments of pulmonary arteries we used 90 stents in 80 pts. Twenty two pts with aortic coarctation and 12 pts with recoarctation underwent stenting of aortic isthmus and we used 34 stents. Six patients with PA type 4 atresia underwent stenting of large aorto-pulmonary collateral vessel. Four patients after Fontan procedure in a modification of total cavapulmonary anastomoses underwent stenting of narrowed intraatrial tunnel. Stenting of stenotic conduit between the right ventricle and pulmonary artery was performed in 6 patients. One patient with stenosis of anastomos Gore-Tex and 1 patient with stenosis of cavapulmonary anastomosis underwent stenting. In one case we carried out the stenting of the atrial septum in patient with mitral valve stenosis. In 30 patients we performed stenting of patent ductus arteriosus. Patients' age varied from 6 hours to 27 years. No serious complications were observed.
Results: All the pts had a good postprocedural result. After stenting the diameter of stenosis increased in average from 5.2 $\pm$2.4 mm to 11.9$\pm$3.1 mm ($p<0.0005$), and systolic pressure gradient fell in average from 54.2$\pm$32.8 to 16.3$\pm$17.1 mmHg ($p<0.0005$). The gradient of systolic pressure in aortic isthmus in patients with coarctation or recoarctation was absent and insignificant in all pts. In patients with large aorto-pulmonary collateral vessel the blood saturation increased from 64% to 78% in first series of cases, and from 70% to 77% in second series of cases after stenting. In the patient after Fontan procedure the symptoms of CHF decreased following stenting. After the stenting procedure of conduits gradient of systolic pressure between the right ventricle and pulmonary artery decreased from 98 mmHg to 35 mmHg. in all cases
Conclusions: stenting is an effective, but technically rather complicated procedure. PA stenting allows to reduce the obstruction and thus prepares patients for the further open heart surgery. It provides better surgical results. Stenting of the aortic coarctation and the recoarctation may serve as an alternative to open heart surgery. The use of stents is also helpful in large aorto-pulmonary collateral vessel, stenosed conduit and intraatrial tunnels.

O14-5
Use of covered stents in the management of native coarctation of the aorta-Diversiry of indications and a special place in developing countries
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Background: The risk of aneurysm formation, dissection, aortic rupture and death following stenting in native CoA, has led to more and more the use of covered stents. Use of covered stents may decrease these risks but does not eliminate them. Along with other indications, covered stents will be especially of value in developing countries where staged dilatation of adds significant cost to the treatment, patient may be lost to the follow up & one off treatment may has be offered.
Objectives: To evaluate the use of covered Cheatham-Platinum (CP) stents in the management of coarctation of the aorta (Co A) in a developing country at a single tertiary care center.
Methods: Covered CP stents were implanted as a primary treatment in at risk patients (May03–Nov06) like complex anatomy with near atretic lesion, long segment coarctation, tortuous arch with marked post stenotic dilatation, transverse arch coarctation, Turner syndrome and as rescue treatment in a previous stent related dissection. Twenty covered CP stents were implanted in 18 procedures on 17 patients. One patient had previous bare stent implantation leading to dissection needing 4 covered stents in 2 procedures.
Results: Mean patient age & weight were (21$\pm$5) years, range 11–46 yrs & (45$\pm$8) kg, range 35–84 kg, respectively. The systolic gradient across the CoA decreased from a mean of 45$\pm$23 mmHg before, to a mean of 4$\pm$3 mmHg after the procedure ($p<0.0001$), and the diameter of the CoA increased from 5.6$\pm$3.9 mm to 16$\pm$3.1 mm ($p<0.0001$). No vascular complications or thromboembolism occurred.
Follow up period was up to 25+15 months (range 2–42 months). One patient had re-dilatation 6 months later. All stents were patent and in good position on CT or MRI performed 3–6 months later. In 33% of the patients antihypertensive medication was decreased and in one patient stopped.
Conclusions: Covered CP stents may be used as the therapy of choice in patients with native coarctation of the aorta with severe and complex coarctation of aorta. They provide a safe cost effective alternative to conventional stenting in patients likely to need serial dilatations and where they may be lost to follow up.

O14-6
Promotion of Growth of True Pulmonary Arteries as a First Step in Staged Repair of Pulmonary Atresia, Ventricular Septal Defect and Major Aorto-Pulmonary Collateral Arteries
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Connection of a hypoplastic true pulmonary artery (PA) to the ascending aorta can be performed as a first step in staged repair of pulmonary atresia with ventricular septal defect and

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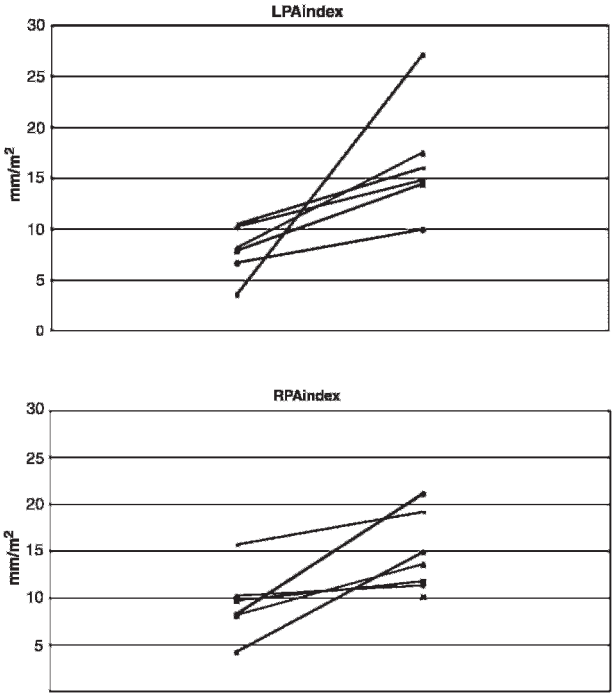
major aorto-pulmonary collateral arteries (PA-VSD-MAPCA) to promote true PA growth. Development of a true PA is prognostically beneficial.
Objective: To determine the early and mid-term growth of true PA's after connection to the ascending aorta.
Methods: We reviewed all angiographic studies of patients with at least two serial studies available, treated in our center in the last 10 years (1996 until 2006) for PA-VSD-MAPCA. Size of right (RPA) or left pulmonary artery (LPA) was measured proximal to the origin of the first branch. For the assessment of growth, the size was indexed to body surface area. Indexed PA-sizes were compared with paired T-test.
Results: Of the 32 cases treated for PA+VSD+MAPCA, 7 underwent connection of true PA's to the aorta as a first step. Mean age at first angiography was 3.8 ± 3.8 months. The last angiography was performed 6.6 ± 5.0 months later. Initial size of RPA and LPA was 2.4 ± 0.8 mm and 2.0 ± 0.5 mm, this corresponded to an indexed size of 9.4 ± 3.7 and 7.8 ± 2.5 mm/m^2. The last measured size for RPA and LPA was 5.0 ± 1.3 mm and 5.4 ± 2.3 mm, respectively. The indexed size of true pulmonary arteries reached 14.6 ± 4.2 mm/m^2 ($p = 0.029$) for RPA and 16.6 ± 5.7 mm/m^2 ($p = 0.035$) for LPA.



Conclusions: Connection of the true PA's to the ascending aorta promotes their growth in PA+VSD+MAPCA and should be considered as an initial step of staged treatment.

O15-1
Persistent Ductus Arteriosus – more than an isolated anomaly
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Background: Persistent ductus arteriosus (PDA) is a common congenital heart disease. Recent animal and genetic studies suggest that PDA can be part of a systemic vascular disease. Clinical case-reports suggest an association with thoracic aorta aneurysms (TAA) or dissection of the aorta (AD). Both TAA and AD are major causes of sudden death.
Aim of the study: We hypothesized that TAA and AD occur more frequently in families of children with PDA.
Methods: Retrospective patient-based study using a questionnaire assessing the prevalence of cardiovascular disease in family members of children with PDA. We took a thorough family history, in which we gathered information about three generations. The statistical analysis was done by a binominal test using SPSS 12.0. The data on the prevalence in the general population was derived from a national population-based study.
Results: 320 patients have been treated for PDA over the last 30 years in our institution. Medical data of 177 families with in total 2511 members were available. The study showed a higher prevalence of anomalies of the aorta in the study group compared to the general population. Aortic pathology was documented in 4/590 = 0.7% of first grade relatives and 29/1339 = 2% of second grade relatives compared to 0.016% in the general population ($p < 0.05$).
In second grade relatives the prevalence of TAA in particular was increased with 0.7% (9/1339) compared to 0.016% in the general population ($p < 0.05$).
By contrast, the prevalence of coronary heart disease was not increased in our study group. It ranged between 0% in probands younger than 45 years and 13.7% among males with a mean age of 66.4 years.
Conclusion: These data provide evidence for an association of PDA and aortic pathology in members of the same family. This suggests that PDA may be a manifestation of a systemic vascular anomaly.

O15-2
Determination of biventricular diastolic compliance and myocardial contractility using Real-Time MRI: a In-Vivo Validation Study
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Background: Load-independent parameters of myocardial contractility and diastolic compliance like the slope of the endsystolic and enddiastolic pressure-volume relation (ESPVR, EDPVR) are fundamental for the evaluation of cardiac function. The conductance catheter (CC) for evaluation of these relation was extensively validated but its technical application is complex and therefore limited in the clinical setting. Aim of this study was the validation of a real-time MRI-method for determination of the EDPVR/ESPVR without geometrical or physiological assumptions.
Materials and Methods: In 6 swine, EDPVR/ESPVR were obtained from biventricular pressure-volume loops derived with CC (gold standard) and MRI methods. MRI was based on real-time measurements of simultaneous biventricular ejection volumes and pressures. Measurements were obtained during balloon occlusion of the inferior vena cava at rest and during Dobutamine.
Results: MRI allowed for fast and reproducible (variation coefficient = 0.4) assessment of the EDPVR/ESPVR. For both ventricles, Bland-Altman analysis showed good agreement between CC and MRI derived parameters, differences were not significant ($p > 0.5$). An increased myocardial ionotropy during Dobutamine was verified in both methods.

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Conclusions: MRI allows to accurately determine both, myocardial contractility and diastolic compliance in the right and left ventricle. The MRI method is, when compared to CC, fast and technically easy to use and therefore applicable in the clinical setting.
This study was supported by Competence Network for Congenital Heart Defects funded by the Federal Ministry of Education und Research (BMBF), FKZ 01G10210.

O15-3
Cardiovascular Findings of Tricyclic Antidepressant Intoxication In Children
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Introduction: Tricyclic antidepressant agents (TCAs) continue to be a leading cause of significant morbidity and mortality in reported poisonings involving pharmaceutical agents. TCAs, particularly amitriptyline, are recognized for their potentially lethal cardiovascular and neurological effects in poisoned patients. Deaths from TCA overdose are usually due to arrhythmias and/or hypotension. Tricyclic antidepressant toxicity is due mainly to the quinidine-like actions of these drugs on cardiac tissues.
Methods: The clinical and laboratory findings of 44 children (19 boys, 25 girls) with amitriptyline intoxication aged between 24 and 192 months who were admitted to our Pediatric Emergency Unit between 2003 and 2006, were retrospectively evaluated. Our purpose was to investigate cardiovascular effects due to amitriptyline intoxication in childhood.
Results: Sixteen out of 44 children took an overdose of amitriptyline for suicide intervention and the remaining took it accidentally. Serum TCA levels varies between 380 and 2000. The most commonly observed clinical findings were unconsciousness, lethargy, and convulsion. Electrocardiographic abnormalities were found in 11 out of 44 children (25%). Sinusal tachycardia was the most common cardiovascular finding. Three of them had long QTc interval, the other three had long PR interval, four of them had prolonged QRS interval, and one of them had nodal rhythm. Serum TCA levels were positively correlated with QRS interval. None of these patients had ST-T changes. Frontal axes were within the normal limits. The children who had ECG findings were older ($p < 0.05$) and had also higher heart rate ($p < 0.05$), serum TCA levels ($p < 0.05$) and had prolonged QRS interval ($p < 0.05$) longer hospital stay ($p < 0.01$) than the children who had no ECG abnormalities All children had been followed-up in PICU and received bicarbonate therapy. Only one child with TCA toxicity who had nodal rhythm, had been given physostigmine. All of the children who had ECG abnormalities had been also given phenytoin infusion. All of the ECG abnormalities which observed in these children, improved within these interventions. None of these children (with or without ECG abnormalities) died.
Conclusion: We consider that electrocardiographic changes should be proposed as a guide to determine the severity of the TCA intoxication and to treat and follow-up the patients with dysrhythmia.

O15-4
Integration of in silico analyses from small pharmacokinetic studies into randomized controlled trials avoid confounding by dose selection in paediatric drug therapy
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Backgr ound and Purpose: Ineffective drug treatment due to underdosing is an ongoing problem in paediatric pharmacotherapy using dosing schedules from adults. Furthermore, if adult dosing regimens are integrated into paediatric randomized controlled trials (RCTs) underdosing may confound the trial result. Therefore age appropriate dosing schedules derived from data analysis and extrapolation by means of in silico models from small size pharmacokinetic paediatric studies performed prior to RCTs might avoid confounding by dose selection.
Methods: Concentration profiles of 25 male and female paediatric patients from a pharmacokinetic study in paediatric patients with congestive heart failure (median age 3.51 years [range 0.07 to 19 years]) treated with the adult doses of oral carvedilol (daily dose 0.7 mg/kg) were analyzed using in silico methods (population pharmacokinetics, POP-PK and simulation analyses) according to the recommendations of the International Conference on Harmonization (ICH) guidelines for conducting paediatric clinical trials. POP-PK and simulations were evaluated using bootstrap analyses, ANOVA and Bonferroni Post Hoc analyses. Decision for the best dosing regimen was based on statistical analysis of the area under the concentration time curve following 0,7, 1, 2, and 3 mg/kg oral carvedilol in infants (28 days to 23 months), children (2 to 11 years) adolescents (12 to 15 years) compared with adults (>18 years).
Results: POP-PK analysis revealed that inter-individual differences in oral clearance and volume of distribution of carvedilol could largely be attributed to size and weight differences. Age showed an additional effect on clearance and volume of distribution. To compensate for age dependent differences daily doses of oral carvedilol (in mg/kg) of 3 were calculated for infants, of 2 for children and of 1 for adolescents for a comparable drug exposition compared to the adult doses.
Conclusion: In silico methods based on small size paediatric pharmacokinetic studies provide age appropriate dose predictions necessary to cope with paediatric needs in pharmacotherapy. Dosing regimens delineated from these results should be integrated into RCTs to avoid confounding by using inadequate dose selection.
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O15-5
Telemedicine as discharge support for families of children with major congenital heart disease.
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Introduction: For parents of children with major congenital heart disease (CHD), the days and weeks following discharge from hospital are a stressful period. A regional service, by its nature, means that many families are distant from the tertiary care centre. Consequently, families may feel isolated and require professional help for issues that arise at times other than their usual clinic review. Home video conferencing not only offers families visual

and audio contact with clinicians but enables visual assessment of the patient.
Methods: This was a randomised control trial comprising three groups. There were two intervention groups – (i) regular video conferences, (ii) regular telephone consultations and one control group receiving usual clinic review. Patients were included if they had major CHD with significant support anticipated after discharge. Qualitative and quantitative data were collected to assess the effect of interventions on patient management, parental anxiety and health care utilisation. A cost analysis was calculated to compare start-up, running costs and hospital admissions.
Results: Of 66 families, 25 were allocated to the video-conferencing group, 22 to the telephone group and 19 to the usual follow up group. The age range of patients was 1 month–3 years. The advice to parents following 26% of telephone consultations was affected by the inability to visually assess patient. The advice to parents following 35% of video conferences was affected by the facility of visual assessment. There was a significant difference between the video conferencing group compared with the telephone consultation group in mean reduction of anxiety ($p < 0.05$). There were fewer patient contact episodes per week in the video-conferencing group (0.4) compared with the telephone group (1.1). There was no significant difference between the three groups in combined start up and running costs and cost of hospital admissions.
Conclusions: Initial results and feedback of this novel study suggest that whilst telephone support is useful, video conferencing has advantages in terms of patient management, parental anxiety. It may also reduce health service utilisation. New video conferencing equipment and internet protocols will reduce costs and should make telemedicine home support an economically viable service.

O15-6

Effects of a 3-month exercise intervention on ambulatory blood pressure and cardiovascular disease risk factors in prepubertal obese children

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Introduction: There is an urgent need for action to strengthen treatment in obese children to pre-empt the spread of the cardiovascular diseases and diabetes. The main purpose of this project was to investigate the effects of a 3-month moderate exercise training program on ambulatory blood pressure and cardiovascular disease risk factors in obese children.
Methods: This was a randomized controlled trial study including 41 prepubertal obese children (22 intervention and 19 controls), aged 6 to 11 years. The intervention consisted of moderate exercise training (ball games, swimming, walking), three times per week (total 180 minutes/week) during 12 weeks. Controls Heart rate was maintained at 55–65% of maximal cardiorespiratory capacity (VO_2peak) using a Polar monitor. Measures included: ambulatory systolic and diastolic systemic blood pressure during 24 hours (ABPM) by Diasys Integra II; body fatness by DXA; VO_2peak by a treamill test; and biological markers of metabolic syndrome (MS): fasting blood lipids (triglycerides, total-, HDL- and LDL-cholesterol), glucose, insulin and CRP levels.
Results: At baseline, all subjects had total body and abdominal fat greater than 25% and a BMI greater than the IOTF cut-off value for overweight. Considering a BP load of 25%, 85% of subjects had systolic hypertension, 49% had diastolic hypertension and 47% had both. The metabolic syndrome was present in 57% of subjects. After 3 months of exercise training (Table 1, intend to treat analysis), 24-hour systolic BP ($P = 0.0079$), percentage of total fat ($P = 0.0001$) and abdominal fat ($P = 0.019$) were significantly decreased, while VO2peak was significantly increased ($P = 0.006$) in the intervention group, compared to the control group. Blood markers did not change significantly.
Conclusion: Hypertension and the metabolic syndrome appear before puberty in a high proportion of obese children. Moderate exercise training during 3 months resulted in decreased ambulatory systolic blood pressure, as well as body fatness and cardiorespiratory fitness. Obese children should be encouraged to participate in moderate physical activities to reduce premature cardiovascular complications.
Funded by the Swiss National Science Foundation and the Research and Development Fund of the University Hospitals of Geneva

Table 1. Physical Characteristics and Ambulatory Blood Pressure Monitoring

Variables	Intervention Group		Control Group	
	Baseline	3 months	Baseline	3 months
Number (female/male)	22 (13/ 9)	22 (13/9)	19 (11/8)	19 (11/8)
Age (years)	9.10 ± 1.5	9.4 ± 1.5	8.8 ± 1.5	9.1 ± 1.5
Height (cm)	140.4 ± 7.9	142.0 ± 8.1	135.3 ± 10.4	137.2 ± 10.3
Body weight (kg)	50.6 ± 11.8	51.6 ± 12.2	47.1 ± 14.8	49.0 ± 15.6
Body mass index ($kg \cdot cm^{-2}$)	25.4 ± 4.6	25.3 ± 4.4	25.2 ± 4.9	25.5 ± 5.2
Body fat DXA (%)	42.5 ± 6.9	41.0 ± 7.0**	42.5 ± 7.2	43.4 ± 7.5
Abdominal fat DXA (%)	50.4 ± 8.0	48.3 ± 8.0*	49.0 ± 8.1	49.8 ± 7.9
VO_2 peak ($ml \cdot kg^{-1} \cdot min^{-1}$)	34.4 ± 5.0	36.3 ± 5.4**	36.3 ± 6.7	34.4 ± 6.3
24h systolic BP (mmHg)	126.1 ± 13.3	120.0 ± 11.5**	125.9 ± 15.8	128.9 ± 18.1
24h diastolic BP (mmHg)	73.2 ± 6.9	70.6 ± 5.4	73.8 ± 8.6	73.5 ± 9.0

MPW-1

Pathogenesis of Pulmonary Arteriovenous Malformations in children with congenital heart disease

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Background: Children with congenital heart disease palliated by Superior Bidirectional Cavopulmonary Anastomosis (SBCPA) develop lung arteriovenous malformations (PAVMs), attributed to lack of hepatic venous blood passing through the lungs. When hepatic venous return is reconnected to the pulmonary circulation by the Fontan procedure, PAVMs regress, suggesting presence of a factor in hepatic venous blood that normally maintains pulmonary vascular integrity. This factor may inhibit PAVM development, or regulate the balance between pro and anti-angiogenic factors. Histology shows vessels to be abnormally large and increased in number within PAVMs, indicating involvement of both dilatory and angiogenic processes. Lung biopsy specimens from SCPA patients show increased Vascular Endothelial Growth Factor (VEGF), but other pro-PAVM factors and the elusive "hepatic factor" remain to be identified.
Aims: To measure plasma levels of potential pro-PAVM factors in children with SCPA and to compare with controls.
Method: A study group of children aged 0–16 with SCPA at risk of developing PAVMs, and a control group of children aged 0–16 with a normal circulation were recruited. During cardiac catheterisation blood was sampled from the hepatic vein,

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superior vena cava, pulmonary artery and pulmonary vein/aorta. Immunoassay was used to measure levels of potential pro-PAVM factors: VEGF, Transforming Growth Factor-ß1 (TGF-ß1), Endoglin and Endothelin-1 (ET-1).
Results: Plasma concentration of VEGF (pg/ml) was significantly increased in the pulmonary circulation in the study group compared to the control group (median values in pulmonary artery 2.52 and 0, pulmonary vein/aorta 4.16 and 0 study and control group, respectively). The patient in the study group with clinically significant PAVMs had the highest values of VEGF. There was no significant difference between plasma concentrations of TGF-ß1 in the study and control groups. However, the patient with PAVMs had some of the highest levels of TGF-ß1; this preliminary data warrants further investigation into TGF-ß1's role in PAVM formation. There was no significant difference in levels of Endoglin and ET-1 between the two groups.
Conclusions: VEGF is a likely pro-PAVM factor in children following BSCPA. Further research into the role of TGF-ß1 in patients with clinically significant PAVMs is necessary to determine its role in PAVM formation.

MPW-2
Cryo-Ablation of Junctional Ectopic Tachycardia
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Background: The congenital form of junctional ectopic tachycardia (JET) is rare. It can be very difficult to achieve control of JET with antiarrhythmic drugs. Radiofrequency ablation has been described but with a risk of atrio-ventricular (AV) nodal block. The purpose of this study was to describe the successful use of cryo-ablation (CRYO) in four patients with JET.
Methods: A retrospective review of patients who had CRYO for treatment of JET. Data included: age, antiarrhythmic therapy, ECG, Holter and electrophysiology study (EPS) results. Success was defined by an absence of JET at follow-up. Standard EPS documented JET prior to CRYO. A 7Fr, 4 or 6-mm-tip Cyrocath Freezor catheter was utilized.
Results: Five patients ranging in age from 2.5 to 15 years underwent CRYO. Prior to procedure they were on an average of 2 medicines (range 1–4) and their tachycardia rate ranged from 120 to 240 beats per minute. The JET focus was localized to the anterior slow pathway/para-Hissian location in all. An average of 3 full Cryo lesions were applied (range 1–4) with each full lesion consisting of 2-minute cycles of freeze-thaw-freeze. Eighty percent of patients had sinus rhythm post-ablation without evidence of JET after CRYO and during follow up Holter monitor at >1 month. First-degree A-V block or transient AV block were noted during failed CRYO so ablation attempts were discontinued.
Conclusion: CRYO of junctional ectopic tachycardia can be successfully achieved with cryo-ablation. No patient in this small group suffered permanent damage to their normal conduction system.

MPW-3
Long-term follow up of bipolar steroid-eluting epicardial pacing leads in children
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Introduction: Cardiovascular abnormalities and vascular size often preclude endovenous pacing leads and necessitate epicardial lead implantation in children. Epicardial leads preserve future venous access and allow placement on the left ventricle. Aim of the study was to evaluate the long-term experience with bipolar steroid-eluting epicardial leads.
Patients and Methods: We prospectively enrolled 114 children, median age 6.2 years (0.0–18.5), and followed up to 12 years (median 3.2). A total of 107 atrial (A) and 132 ventricular (V) bipolar steroid-eluting epicardial leads were implanted (Medtronic CapSure Epi 10366 or 4968). Telemetry data were obtained at implant and semi-annual visits. Pacing thresholds were calculated for a standard value of 0.5ms pulse duration. For analysis, follow up was truncated at 8 years. Children with left or right atrial (LA/RA) and ventricular (LV/RV) pacing and with prior or no surgery were compared. Data are given as median.
Results: Pacing thresholds remained below 1.2 Volt (A: 0.66–1.12 Volt, V: 0.83–1.18 Volt). Neither difference was found for pacing thresholds between pacing sites (LA/RA: 0.82 versus 0.74 Volt, $p = 0.85$; LV/RV: 0.96 versus 0.94 Volt, $p = 0.65$), nor prior or no surgery (A: 0.87 versus 0.67 Volt, $p = 0.56$; V: 1.01 versus 0.83 Volt, $p = 0.21$). Sensing demonstrated a superior trend of LV-leads (LV/RV: 11.2 versus 7.7 mVolt, $p = 0.002$). Kaplan-Meier estimates of freedom from lead failure at 2, 5 years was 99, 94% for A- and 96, 85% for V-leads. Freedom from lead failure did not differ between pacing sites (LA/RA: $p = 0.37$, LV/RV: $p = 0.92$).
Conclusions: Epicardial leads provide low atrial and ventricular pacing thresholds up to 12 years of follow up, independently of the pacing site or prior cardiac surgery. Adequate atrial and ventricular sensing with a superior trend of LV-leads was observed. Estimated freedom from lead failure showed favourable lead survival in the growing patients. These encouraging results consolidate the epicardial approach as the first choice in children with permanent pacing and lifelong pacemaker-dependency.

MPW-4
Short term follow up of implantable Cardioverter Defibrillator Systems using Epicardial and Pleural Electrodes in Pediatric Patients
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Introduction: The optimal electrode and device placement for implantable cardioverter defibrillator (ICD) therapy in children is still controversial. We describe our initial experience after epicardial and pleural electrode insertion with abdominal or intrathoracic ICD placement.
Patients and Methods: An ICD system was implanted in 8 children (median age 11.6 years) due to hypertrophic cardiomyopathy ($n = 6$), long QT syndrome ($n = 1$) and Brugada syndrom ($n = 1$). After muscle sparing left lateral thoracotomy, a tunnel was created along the third intercostal space for insertion of the defibrillation electrode. Pacing and sensing electrodes were secured on the left atrium and ventricle. The device was placed in the rectus sheath ($n = 5$) or, to optimize the electrical field, in a horizontal position

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within the diaphragm underneath the right ventricle (n = 3). Three patients underwent concomitant cardiac surgery. ICD follow-up data were obtained at 1, 3, and thereafter at 3–6 months intervals.
Results: No implant complications were observed. Median defibrillation threshold at implant was 20 Joules (range 10–25), even when testing was done immediately after cardiopulmonary bypass.
During a maximum follow up of 39 months (median 21), 3 appropriate and successful ICD discharges were seen in 1 patient. Two inappropriate shocks were documented due to sinus tachycardia and T-wave oversensing. Two system revisions were required due to dislodgement of the defibrillation coil electrode in the early learning curve and an inner isolation defect of a ventricular lead 17 months post-implant. Furthermore, one atrial lead fractured occurred 25 months post-implant. Twelve months after implantation, routine defibrillation threshold testing was done in 4 patients, and demonstrated stable thresholds.
Conclusions: Short-term experience demonstrates that an ICD system with epicardial sensing and pacing leads and a subpleural placement of the defibrillation electrode is feasible and safe. The intra-thoracic position of the device results in a safe and protected position. Long-term follow-up data are needed to promote this alternative ICD implant technique for children and patients with congenital heart disease with limited venous access.

MPW-5
Congenital heart block not associated with antiRo/SSA antibodies
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Purpose: Isolated congenital complete heart block (CHB) is usually associated with maternal anti-Ro/SSA antibodies. We report the largest case series of CHB not associated with anti-Ro/SSA antibodies. Aim of our study was to assess the prevalence of anti-Ro/SSA and/or La/SSB negative CHB and its clinical course.
Methods: Setting: 2 tertiary referral centers. We observed 46 fetuses with isolated advanced fetal atrioventricular (AV) block detected during pregnancy. Maternal anti-Ro/SSA and La/SSB antibodies were detected by counterimmunoelectrophoresis, ELISA and line blot.
Results: In 36 cases the mothers were anti-Ro/SSA positive (78.2%) and in 10 they were negative (21.8%).
Ro/SSA POSITIVE INFANTS. M/F ratio was 10/23. 10/36 infants died (27.8%): 3 for intentional abortion, 2 suddenly in utero, and 5 soon after birth (4 for intractable heart failure and 1 suddenly); another developed severe dilated cardiomyopathy one year after birth. Mean gestational age at detection of CHB was 24.7 wk. Almost all infants had a complete AV block, two incomplete AV blocks reverted to sinus rhythm after high dose dexamethasone *in utero*. The mean heart rate at birth was 60 bpm; 29/31 babies were paced, at different ages.
Ro/SSA NEGATIVE INFANTS. No mothers had a connective tissue disease. M/F ratio was 1/1. 3/10 infants died after birth only one for cardiac causes (heart failure) . Two infants had a stable III degree AV block at birth; 5 had II degree AV block, that progressed to a high degree block in 4. In 2 patients AV block reverted to normal sinus rhythm at birth. Mean gestational age at detection of CHB was 29.6 wk and mean heart rate at birth was 78.5 bpm; 6/10 infants were paced.
Conclusions: 21.8% of isolated CHB were anti-Ro/SSA negative. Their mothers had no connective tissue disorders. Compared to anti-Ro/SSA positive CHB, these blocks were detected later during pregnancy,were less stable, and have a slower and more benign evolution.

MPW-6
Assessment of cardiac function in severe twin to twin transfusion syndrome – multicenter study
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Objective: To investigate cardiac function in both twins in course of TTTS before and after treatment.
Methods: In a retrospective study 28 pairs of twins with TTTS were evaluated, between 16 and 29 (mean 21) at the first echo exam. Cardiovascular profile score (CVPS) was used for evaluation of the cardiac function and peripheral circulation. CVPS at the first and final examination before delivery was compared in both twins. Fetal outcome was assessed.
Results: The median CVPS for recipients at the first assessment was from 3 to 9, and from 8 to 10 for donors. CVPS was above 7.0 for survived recipients. CVPS was higher for donors, however and it was not correlation between CVPS and survival for donors. Cardiac function was impaired in all recipients: tricuspid regurgitation (100%), mild cardiomegaly (75%) and myocardial hypertrophy (56%) were the most frequent findings. Functional pulmonary atresia in four cases and supravalvar pulmonary stenosis in two was diagnosed. Cardiac function was abnormal in three donors twin due to CHF after laser therapy. There was functional pulmonary atresia in two of them. In one, premature ductal occlusion was diagnosed. In utero treatment included: serial amnioreduction, laser therapy, and digoxin. Overall survival rate was 53% for donors and 47% for recipients 47%.
Conclusions: 1.The CVPS is useful in the surveillance of recipients for prediction of the presence of congestive heart failure and fetal outcome. 2. Cardiac function was impaired in all recipients whereas donors showed peripheral flows abnormalities with similar mortality rate in both groups. 3. Impaired cardiac function was seen in some of donors after laser therapy, what is new finding in such clinical condition.

MPW-7
Maternal Ingestion of Polyphenol-rich Common Beverages is Associated to Higher Fetal Ductus Arteriosus flow Velocities and Larger Right Ventricular size in Normal Pregnancies
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Background: We have already demonstrated that maternal consumption of green tea, mate tea and grape juice, beverages with high polyphenol compounds (3-gallate-gallocatechin and resveratrol) cause fetal ductal constriction, tiggered by their anti-inflammatory effects as a result of cyclooxygenase-2 inhibition and prostaglandins. This study test the hypothesis that in normal pregnancies maternal ingestion of polyphenol-rich common

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beverages interferes with fetal ductal flow velocities and with the size of the right ventricle.
Methods: A prospective analysis of 140 third trimester fetuses from normal mothers was carried out, determining systolic and diastolic ductal flow velocities and right to left ventricular dimensions ratio. Mean maternal age was 30.3 ± 5.4 years (15 to 43 years) and mean gestational age 28.4 ± 3.1 weeks (23–38 weeks). A questionnaire was applied about maternal consumption of polyphenol-rich substances during pregnancy (herbal teas, mate tea and grape derivatives).
Results: The group of 100 fetuses whose mothers declared to have used herbal teas or grape juice derivatives showed higher mean systolic (0.96 ± 0.23 m/s) and diastolic (0.17 ± 0.05 m/s) velocities, as well as higher RV/LV ratios (1.23 ± 0.23) than the group of 40 fetuses whose mothers have not utilized these substances (mean systolic velocity: 0.61 ± 0.18 m/s, P = 0.000, mean diastolic velocity: 0.11 ± 0.04 m/s, P = 0.011, and mean RV/LV ratio: 0.94 ± 0.14, P = 0.000). Systolic ductal velocities were correlated with RV/LV ratio (r = 0.64, P = 0.000).There was association between maternal comsumption of herbal teas and grape derivatives and fetal systolic ductal velocities >0.85 m/s (P = 0.000, relative risk = 8.26, 2.75–24.81), diastolic velocities >0.15 m/s (P = 0.000, relative risk = 2.57, 95% CI 1.41–4.69) and RV/LV dimension ratios >1.1 (P = 0.000, relative risk = 27.6, 95% CI 3.96–192.01), independently of gestational age.
Conclusion: Ductal flow velocities are higher and right to left ventricular dimension ratio is larger in fetuses exposed to maternal ingestion of polyphenol-rich substances than in those not exposed. It seems clear that ductal flow response to consumption of polyphenols during pregnancy is not a categorical parameter, but rather a continuous dose-dependent variable.

MPW-8
Fetal Diagnosis of Cardiac Rhabdomyoma, its Management and Outcome
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Objectives: This study analyses the long term cardiac and neurological outcome of patients with cardiac rhabdomyoma (CR) in order to allow comprehensive prenatal counselling.
Background: Because of the relative rarity of the disease, there is paucity of data concerning the outcome of patients with CR.
Methods: A retrospective study including all cases with echocardiographic diagnosis of CR encountered between April 1986–August 2006.
Results: Of 24 CR patients identified, 7 were diagnosed in-utero at a gestational age (GA) between 28–35 weeks and 17 postnatally between 10 days–5 years. 14 had multiple CR and 10 had one/two CR. The CR were situated predominantly in the LV (70%), RV (52%) and IVS (48%) and to a lesser extent in the atria (13%) and pericardium (4%). Follow-up echocardiography in 18 showed complete postnatal regression of CR in 3, partial regression in 13 and no change in 2. Cardiac complications were encountered in 5 patients, 1 with WPW syndrome and SVT requiring anti-arrhythmic therapy, 1 with sub-aortic obstruction needing surgical intervention and 3 with occasional bouts of paroxysmal SVT. Long-term follow-up revealed tuberous sclerosis of Bourneville (TSB) as definite diagnosis in 22, complicated by epilepsy in 16 and developmental delay in 14.
Conclusions: CR generally regresses after birth and after the high risk perinatal period cardiac related problems are rare. The relatively poor neurodevelopmental outcome of the almost always associated TSB however should form a dominating aspect of the prenatal counselling of parents whose fetuses are diagnosed with this rare disease.

MPW-9
Missed Diagnoses in Antenatal Screening for Congenital Heart Disease: Implications for Training
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Introduction: Congenital heart disease (CHD) is the most common severe fetal abnormality. 90% of major CHD occurs in low risk pregnancies. Screening takes place during the anomaly scan at 20 weeks gestation. The AEPC recommend that, as a minimum standard, a 4 chamber view of the heart should be obtained during anomaly screening. In 60% of fetuses with major CHD the defect can be seen on the 4 chamber view. Addition of outflow tract views can raise antenatal detection of major CHD to over 90%. This study aims to ascertain which types of CHD are missed antenatally in Northern Ireland (NI) and compare detection of 4 chamber lesions with outflow tract lesions.
Methods: All cases of major CHD in NI between 1st January 2003 and 31st December 2005 were identified using the regional CHD database, fetal cardiology clinic and postmortem records. Cases were classified according to whether they could be detected on a 4 chamber view, or required outflow tract visualisation to identify a defect. 77 of the 88 obstetric radiographers in NI (88%) attended fetal cardiology training. All were given an anonymous questionnaire regarding their anomaly scanning practice.
Results: There were 198 cases of major CHD in NI during the 3 year period. 24.2% were diagnosed antenatally. 65.1% of these were considered identifiable by antenatal scanning of the 4 chamber view. 34.9% would have required visualisation of the outflow tracts for detection. 34.1% of 4 chamber lesions received an antenatal diagnosis. This was the case for only 5.8% of outflow tract defects. 72 radiographers (94%) completed questionnaires. 43% of respondents limit the assessment to the 4 chamber view. 36% attempt to view one outlet, 21% look for both outlets.
Conclusions: Antenatal diagnosis of major CHD in NI is suboptimal. Diagnosis of lesions affecting outflow tracts is significantly lower than that of lesions visible on the 4 chamber view. Obstetric radiographers require further training in the assessment of the fetal heart (in particular in scanning the outflow tracts). A regional training programme has been implemented. Screening of the fetal heart should include assessment of outflow tracts in all cases.

MPW-10
Polish National Database for Fetal Cardiology at www.orpkp.pl
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The National Database for Fetal Cardiac Pathology was established in 2004. There are three levels of fetal cardiac centers based on a number of diagnosed cases with major cardiac problems per year: A-minimum 10; B–minimum 50; C-minimum 100.
Each fetal record is subdivided into following forms: center of diagnosis, patient's coded ID, name of referring physician, name

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of fetal cardiac consultant, fetal cardiac diagnosis, extra cardiac diagnosis, therapy (in selected cases), delivery, postnatal follow-up. 18 centers cooperated: 3C, 3B, 7A and other who submitted less than 10 fetuses. Each of three major fetal cardiac centers type C is obliged (in shifts) to verify the new records.
In June 2004 there were 457 records, in November 2005 – 655, in December 2006 – 1125. 87% of all records were fulfilled by 4 centers (3 type C and one type B).
We are using AEPC codes. At the beginning we used long list, which was too detailed for the analysis purpose. It was changed into modified short list, but still this list provides too many codes for similar abnormalities.
We made subdivision for CHD as: critical, severe and benign. The most common defect was HLHS (6.3%). We were also interested in severe fetal cardiac arrhythmias, such as SVT or complete heart block. Premature atrial contractions were not collected in this database. On-line reports are available, for instance: number of fetuses with cardiac problems presented with polyhydramnion, oligohydramnion, extracardiac anomalies, abnormal karyotype. Out of 470 completed records we learned that 50% of deliveries were by cesarean sections, which was unexpected finding.
Based on the data from the database we invented clear definition of two Certifications for physicians illegible for fetal cardiac screening and for advanced fetal echocardiography examination.
In coming year 2007 our major plans are: 1) changes of AEPC cardiac codes to more adjustable for fetal cardiology, 2) collect a freeze and/or cine-loop in digital form for diagnostic and educational purpose.
Conclusions: National database base for fetal cardiac problems is full of precious information allowing to plan future of fetal cardiology and improve organization of the cardiac perinatal care.

MPW-11
Novel Therapeutic Strategy for Eisenmenger Patients in Childhood
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Introduction: In patients with congenital intracardiac shunting defects, corrective surgery may be unfeasible due to elevated pulmonary vascular resistance (PVR). A pronounced vasodilatory response upon pharmacological testing ("responder") might indicate a therapeutic option to achieve reversibility of pulmonary vasculopathy. To favour pulmonary vascular remodelling with the objective of regaining operability we performed a hybrid therapy consisting of surgical pulmonary artery banding (PAB) and combined targeted medical therapy including bosentan (Tracleer®) and sildenafil (Revatio®) in 4 children with Eisenmenger physiology due to simple intracardiac shunting defects.
Methods: 4 children (aged 3.4, 3.6, 4.4 and 9.7 years) with ventricular septal defect in 3 and atrioventricular septal defect in 1 underwent cardiac catheterization showing systemic levels of PVR but preserved pulmonary vasodilatory reserve. Surgical PAB was performed and followed by the beginning of a medical treatment including bosentan and sildenafil. During follow-up, pulmonary artery pressures (PAP) and right ventricular systolic pressures (RVSP) were obtained by cardiac catheterization to evaluate the progression of disease and response to therapy. Changes in hemodynamic variables were evaluated using student t-test.
Results: During a mean treatment period of 4.5 years (3.4–6.1 years), clinical stability remained in all 4 patients. No complications following surgical intervention, or adverse events related to specific pulmonary vasodilative therapy were observed. Mean transcutaneous oxygen saturation decreased slightly from $91.8 \pm 7\%$ to $90.0 \pm 3\%$. Mean PAP decreased significantly from 73 ± 7 mmHg before therapy to 49 ± 14 mmHg (most recent value); $p < 0.05$. Mean RVSP also showed a significant reduction from an initial value of 103 ± 5 mmHg to 88 ± 11 mmHg; $p < 0.05$. For these two hemodynamic variables, a descending trend towards normal values could be monitored over time.
Conclusions: Combination of pulmonary artery banding with specific pulmonary vasodilative therapy caused significant reduction of pulmonary artery and right ventricular systolic pressure in these 4 patients. We suggest that in patients with inoperable intracardiac shunting defects and severe pulmonary vasculopathy this novel approach may be a therapeutic option which is worth being evaluated for the potential of regaining operability and thus avoiding transplantation.

MPW-12
Pulmonary Blood Flow in Eisenmenger Patients Measured by Inert Gas Rebreathing
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Introduction: In patients with Eisenmenger syndrome, hyperplasia and hypertrophy of the pulmonary arterial smooth muscle cells, and pulmonary vasoconstriction all cause a reduction of pulmonary blood flow (PBF) and thus impairment of exercise capacity and quality of life. Improving lung perfusion therefore is a major target of pharmacological therapy in these patients. Monitoring therapeutic success remains a challenge because changes in hemodynamic parameters like pulmonary artery pressure, pulmonary vascular resistance and PBF are difficult and mostly only invasively to obtain. This study was performed to assess the feasibility of measuring PBF noninvasively using a new device (INNOCOR®).
Methods: 15 patients with Eisenmenger syndrome, median age 30 years (17–44 years), and 8 healthy untreated volunteers, median age 27 years (12–45 years) were studied. Measurements of PBF ($l/min^{\star}m^2$) were performed at baseline and 24 weeks after initiation of pulmonary vasodilative therapy with bosentan using an inert gas rebreathing technique (INNOCOR®). The principle of this method is to breathe a gas mixture containing one blood soluble (nitrous oxide, N_2O) and one blood insoluble compound (sulphuric hexafluoride, SF_6). PBF is directly proportional to the rate of washout of N_2O which is measured continuously by a gas analyzer.
Results: Baseline measurements showed a highly significant difference of median PBF values between the volunteer group and the Eisenmenger patient group (3.46 ± 0.94 $l/min^{\star}m^2$ versus 1.76 ± 0.39 $l/min^{\star}m^2$; $p < 0.001$). No differences in PBF could be demonstrated in the small patient group after 24 weeks of bosentan treatment; in contrast, they showed both a significant reduction of pulmonary vascular resistance measured during cardiac catheterization and a remarkable improvement of exercise capacity.
Conclusions: The inert gas rebreathing technique was used as an easy, safe and quick method to measure PBF noninvasively and confirmed severely reduced PBF in the Eisenmenger patient group. Further studies are warranted and under way to answer the question whether this method is sensitive enough to detect the probably small changes of PBF caused by specific pulmonary vasodilative therapy and wether these correlate with the changes of clinical status and exercise capacity in these patients.

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MPW-13

Cardiac function analysis with MRI: Assessment of measurement accuracy in a prospective multicenter study

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Background and aim: MR volumetry is considered the gold standard to assess cardiac function but interobserver variability remains a practical limitation. We sought to assess the impact of semi-automatic post-processing software and consensus criteria for image reading to reduce interobserver and interexamination variability (IOV, IEV).

Materials and Methods: We designed a prospective multicenter study involving patients with congenital heart disease (Fallot, DORV, n = 45) and controls (n = 15). All subjects were studied at 1.5 T scanners (Philips) using well defined imaging protocols acquiring: (1) LV and RV enddiastolic/systolic volumes (EDV, ESV) with multislice SSFP in a short-axis and transversal plane; (2) muscle mass of interventricular septum, LV and RV freewall. MRI data was analyzed by customized software using semi-automated border detection by threshhold adjustments. Images were acquired at three centers. Data were blinded and analysed by three experienced independent observers before and after informed consent about standardized guidelines for image analysis.

Results: Interexamination variability was low (<4%) in all studies and there was no systematic measurement error for either operator ($p > 0.62$). However, without informed consent there was significant IOV with a maximum of 18% for RV-EDV measured in short-axis plane in patients and a minimum of 6% for LV-ESV in controls. After informed consent variability decreased significantly ($p < 0.01$) to 6% and 4%, respectively. Bland-Altman test showed agreement between measurements acquired in short-axis and transversal planes, however, IOV was significantly higher ($8 \pm 7\%$, $p < 0.05$) in short-axis when compared to analysis done in transversal planes.

Conclusions: Even in the presence of high-quality SSFP source images for MR volumetry with low IEV, consensus guidelines and training for image analysis were essential to reduce IOV to overall less than 6%. This is a fundamental requirement to enable for relaible quantification of cardiac function in patient follow-up and clinical research. Transverse-plane volumetry is less prone to IOV than short-axis volumetry.

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MPW-14

Effect of high-dose betablocker therapy on exercise capacity in patients with hypertrophic cardiomyopathy

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Introduction: Hypertrophic cardiomyopathy (HCM) is a common medical cause of sudden death during exercise in childhood and adolescence. High-dose beta-blocker therapy (HDBB; = propranolol equivalent doses of >4.5mg/kg) reduces risk of sudden death, but its use in low-risk patients is controversial, as there is a perception that exercise ability will be drastically curtailed. However in HCM impaired diastolic function is a major limiting factor for exercise capacity, and HDBB improves diastolic function in HCM. In a randomised prospective trial of HDBB in low-risk HCM-patients we are therefore studying exercise capacity.

Methods: 11 HCM patients have >one year follow-up, 6 in HDBB group, median age 12 years (range 9–15 years), and 5 Controls, median age 13 (range 7–24 years), with no medical treatment. Both groups received advice concerning life-style modifications according to American Heart Association Guidelines. Before study entry, and after one year they underwent incremental bicycle exercise tests (Monark ergomedic 839E) with a ramp protocol starting at 1W/kg, with 10W increments each minute. During the test we monitored 12 lead ECG, blood pressure, and heart rate every minute.

Results: Maximum exercise capacity in W/kg in the HDBB-group before treatment, median 3.0W/kg (range 2.1–3.8) was not different from Controls, median 2.6W/kg (range 2.1–3.3: $p = 0.08$). After one year of therapy the exercise capacity of HDBB-group, median 2.7W/kg (range 2.3–3.0) was not significantly reduced compared with pre-treatment values ($p = 0.11$) and no different from the Control group, median 2.7W/kg (range 2.1–2.9); ($p = 0.99$); within normal range for age. In fact 4/6 in HDBB-group improved their total workload (median change +8W), whereas 0/5 Controls did (median change −5W). Maximal heart rate on exercise was reduced by HDBB from median 182b/min (range 159–190) to a median 128b/min (−30%, range 118–131) $p = 0.03$, whereas it was unchanged in Controls, initially median 187b/min (range 168–198) after one year median 186b/min (range 150–188). No HDBB-patient developed pathological blood pressure fall during or after exercise on treatment.

Conclusions: In spite of a 30% reduction in maximal heart rate on exercise, HCM-patients on HDBB-therapy do not show a significantly reduced exercise capacity, probably because improvement in diastolic filling increases the stroke volume to compensate.

MPW-15

Selection of high risk for death subgroups at presentation by classification tree analysis in infantile idiopathic dilated cardiomyopathy

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Background: Infantile idiopathic dilated cardiomyopathy has high mortality. The final treatment is heart transplantation and in the future, stem cell transplantation. Nevertheless, there is not criterion to anticipate the death at presentation.

Objective: Select subgroups of children with high risk of death by idiopathic dilated cardiomyopathy using classification tree analysis.

Patients and Methods: This is a retrospective study of 179 children with idiopathic dilated cardiomyopathy (45 deaths). It was analyzed 36 clinical parameters and 25 from complementary exams. To build the tree it was employed CART algorithm, with selection by GINI index and prune by cost-complexity, aiming to maximize probability reason.

Results: From clinical and laboratorial data, it was built a tree with eight branches and seven nodes corresponding to seven variables,

selecting three high-risk subgroups for death at presentation: a) children in functional class IV and heart rate less than 133 bpm; b) children in functional class IV, heart rate greater than 133 bpm, mitral regurgitation grade 3–4, cardiothoracic ratio greater than 0.62 and ascites and c) children in functional class IV, heart rate greater than 133 bpm, mitral regurgitation grade 3–4, cardiothoracic ratio greater than 0.62 without ascites, but left ventricle short fraction less than 0.12 and left ventricle mass/body surface area grater than 177.3 g/m^2.
Conclusion: From these select parameters, it is possible anticipate risk of death at presentation. If therapeutic response is not adequate, then heart transplantation and perhaps in the future, stem cell transplantation, should be indicate, with setting the child in waiting list.

MPW-16
Long-term follow up of neoaortic regurgitation (AR) after the arterial switch operation for transposition of the great arteries
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Objective: Anatomical correction (ASO) is the therapy of choice for patients with simple and complex form of TGA. In our institution between July 1991 and January 2007 over 460 children underwent ASO.
The Aim of the Study was to determine the outcome of neoaortic valve after arterial switch operation. We sought to identify predictors of AR.
Methods: Included in the study were 180 patients who underwent ASO from 1992 through 2000, in the age of 5 to 16 years, in four groups: I-112 children with TGA+IVS, II-46 pts with TGA+VSD, III-11 pts with TGA+AAA, IV-11 pts after two stage op. (previous PAB). We followed pts with echocardiography at 5, 7, 9, 11, 13 years after ASO. The AR was graded as 0/4-none, 1/4-trivial, 2/4-mild, 3/4-moderate, 4/4-severe. Statistical analysis was performed using Statistica 6.0. A p-value less than 0.05 was considered to be statistically significant.
Results: We found 46 pts with AR grade1/4 (26%), 12 grade 2/4 (7%) 5 years after ASO. Probability of freedom from AR was in total group 67%, 38% (r2-0,77; p < 0,05) and in I group 72%, 40% (r2-0,7; p < 0,05) at 5 and 13 years respectively. AR progressively increases with follow-up: in the I group: 1/4-23%, 41%, 51%,46%, 50% (r2-0.7, p < 0.05); 2/4-5%, 8%, 3%, 8%, 10%; in the II group: 1/4-24%, 14%, 0%, 8%; 2/4-6%, 14%, 17%; in the III group: 1/4-27%, 50%, 50%, 50%, 2/4-9%, 25%, 25%, 50%; in the IV group: 1/4-55%, 70%, 2/4-18% after 5, 7, 9, 11, 13 years respectively. One patient (from the IV group) developed after 11 years severe AR and required aortic valve replacement. 6 pts among 21 with at least 2/4 AR (29%), vs. 9 pts among 159 freedom from AR or 1/4 AR (6%) received ASO later then in 2 month of life. In that group of 15 pts there were 9 pts from the IV group. The existence of VSD itself was not found to be significant risk factors in our series.
Conclusions: Aortic regurgitation (AR) continue to develope over time after ASO.
Previous PAB and older age at time of ASO were significant risk factors.

MPW-17
Role of Cardiac Troponin T in Clinically Suspected Acute Myocarditis in Children
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Background: Endomyocardial biopsy has historically been the reference standard in diagnosing myocarditis. As inflammation of the myocardium may be patchy & easily missed, the sensitivity of EMB remains low. Moreover, the risk of cardiac perforation with EMB is increased in young children. In our part of the world where incidence of myocarditis is high, it has significant cost implications as well. Cardiac Troponin T is a sensitive marker of myocardial injury and is known to be raised in acute myocarditis.
Objective: To describe the serum troponin T levels in children presenting with acute myocarditis.
Design: Descriptive study performed at a tertiary care paediatric cardiology center from July 05 to Dec 05.
Material and Methods: All consecutive patients above 1 month of age but age less than 14 years, admitted with clinically suspected acute myocarditis were included by convenience sampling. Diagnostic criteria were (1) the presence of severe and acute heart failure; (2) left ventricular dysfunction on echocardiography; (3) recent history of viral illness; and (4) no history of cardiomyopathy. Complete investigations were performed to rule out congenital heart malformations (for example, coronary artery anomalies). Patients with metabolic perturbations, exposure to cardiotoxic agents, chronic primitive arrhythmia and chronic bacterial sepsis were excluded. Serum Troponin T (Quantitative) level were performed. The cut off level of .052 ng/ml was taken as cutoff point to diagnosis acute myocarditis.
Results: Of twenty-five patients (n = 25), male to female ratio was 2:3 (10/15). Mean age was 23.7+1 SD 4.9 months (range 4 months –12 years). Mean ejection fraction (EF) was 36% (range 18%–45%). 17/25 had acute fulminant presentation. Mean Troponin T level was 0.231 ng/ml (range .007–1.06) with median of 0.119 ng/ml. Considering 0.052 ng/ml as positive cut off value, 15/25 (60%) had positive Troponin T test. All these 15 children had acute fulminant myocarditis. Mean troponin T levels (quantitative) was analyzed and correlated with ejection fraction.
Conclusions: Cardiac serum troponin T is raised in all children with acute fulminant myocarditis in our set up. There is no correlation between troponin T and ejection fraction. More studies need to be done to calculate specificity.

MPW-18
Long-term outcome of pediatric patients with biopsy-proved myocarditis: comparison with late stage Kawasaki disease
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Purpose: Myocarditis is one of complications of Kawasaki disese (KD) and may persist in the late stage. The study objectives were to assess the long-term outcome of patients with biopsy-proved lymphocytic myocarditis (Dallas criteria), and to compare the outcome of these patients with that of patients with late stage Kawasaki disease (KD). To clarify the myocardial damages in chronic stage of KD, biochemical markers and dual isotope

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single photon emission computed tomography (SPECT) and endomyocardial biopsy (EMB) were compared in KD.
Patients and method: They included 6 patients with biopsy-proved myocarditis and 28 KD with coronary artery lesions (CAL). Ages at onset of diseases ranged 2 to 18 years. Biochemical markers were high-sensitive C-reactive protein, myoglobin, Creatin Kinase MB, troponin T, heart-type fatty acid binding protein, ANP and BNP. Histopathology of endomyocardial biopsy was evaluated with histomorphometric method by computer assistance. A defect score for SPECT images was interpreted as normal: 0, mildly decreased: 1, moderately or severely decreased: 2, complete defect: 3.
Results: The patients with biopsy-proved myocarditis presented sudden onset syncope in two, myocardial infarction-like symptoms in two and acute DCM in one. Three of those showed complete improvement, one was required PMI 6 years later due to complete AV block. KD patients were all free from cardiac symptoms at late stage. Myocardial changes on EMB at chronic stage of KD showed various histopathological findings such as fibrosis, degeneration, disarray and inflammatory cell infiltration in the patients with CAL, including suggested chronic myocarditis. Accumulation of myelin bodies in the myocytes, disarray of myofibrils, vacuoles and microangiopathy were found as ultrastractural changes.
TL/BMIPP discrepancy was found in KD with CAL. In the cases with TL/BMIPP discrepancy, ultrastructual changes revealed massive myelin bodies. Conclusion: We found that the long-term outcome of patients with biopsy-proved myocarditis was not always benign and the patient with a clinically silent myocarditis during follow-up developed complete AV block. Discrepancy of dual SPECT image suggested that myocardial changes in long-standing Kawasaki disease might reflect not only myocardial ischemia but also disordered myocardial fatty acid metabolism following myocarditis. Further studies are needed to clarify residual myocardial damages following KD.

MPW-19

Prevention of Respiratory Syncytial Virus Infection in Young Children With Heart Disease – A National Multicentric Study in a Mediterranean Country

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Introduction: Bronchiolitis due to the respiratory syncytial virus (RSV) is a major cause of morbidity and mortality in children below two years of age and an important cause of hospital admission during wintertime. Children with significant congenital and acquired heart disease represent a high-risk group for RSV infection with increased morbidity and mortality. Several studies have demonstrated the efficacy and safety of Palivizumab prophylaxis. Palivizumab has been shown to decrease the severity and incidence of RSV infection in this group of patients.
Objective: To assess the safety, tolerance and efficacy of Palivizumab prophylaxis in children with hemodynamically significant heart disease in a Mediterranean country.
Methods: Prospective paired control multicentre study in consecutive children less than two years of age with major heart disease, for two RSV seasons (2005–2006). RSV hospitalizations and length of hospital stay were used to evaluate efficacy. Children were followed for a period of 210 days. Descriptive statistics was used. Comparative analysis included Wilcoxon ranking and McNemar test (at 0.05).
Results: 696 children were included (348 palivizumab group; 348 control group); there were no differences in demographic, heart disease and risk factors in both groups. 50% were male, 46% had cyanotic congenital heart disease. RSV hospitalization was significantly higher in the control group (13% vs. 4%, $p < 0.001$). In the palivizumab group with RSV hospitalization ($n = 13$; 4%) 38.5% had cyanotic heart disease and 61.5% congestive heart failure. The length of hospital stay was 11 days for the palivizumab group and 20 days for controls. There were no cases of mechanical ventilation in palivizumab group. During the study, we observed 8 adverse events in 6 children, but only three were drug related. No mortality was registered in both groups.
Conclusions: In the studied population, palivizumab was safe, well-tolerated and effective in reducing clinical impairment and hospital admissions in children less than 2 years old with hemodynamically significant heart disease. Although our data are similar to others previously published to our knowledge this steady is the first one realized in a Mediterranean country with data of two RSV seasons.

MPW-20

The Role of Telemedicine in the Remote Diagnosis of Congenital Heart Disease

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Objectives: To evaluate the role of telemedicine in the detection of congenital heart disease (CHD) by assessing the diagnostic accuracy, impact on patient management and cost implications of echocardiograms transmitted across Integrated Systems Digital Network (ISDN) lines with expert guidance.
Methods: Prospective study of consecutive patients in three district general hospitals (DGH) in whom significant CHD was suspected by the attending paediatrician. An initial echocardiogram by the paediatrician was compared with the transmitted echocardiogram with a third "hands-on" echocardiogram as gold standard. The images were transmitted to the regional paediatric cardiology unit across ISDN 6 lines with live guidance and interpretation by a paediatric cardiologist.
Results: Over an eight year period echocardiograms were transmitted on 124 infants. Five transmitted scans were inadequate. "Hands-on" echocardiograms were performed on 109 of the remaining 119 tele-echocardiograms (92%). Major CHD was diagnosed in 39/109 infants (36%) and minor CHD 46 (42%). The tele-echo diagnosis was accurate in 96% cases (Kappa score 0.89). There were 4 diagnostic errors. Transfer to the regional unit was avoided in 89/119 patients (78%). Each DGH incurred a net cost during the study. A large proportion of the cost (40%) was purchase of equipment.
Discussion: This is the second largest European study to date of this application of telemedicine. The high follow up rate allows confident validation of this process. CHD is accurately diagnosed by real time transmission of echocardiograms performed by paediatricians, with no formal training, under live guidance and interpretation by a paediatric cardiologist. Telemedicine affects the management of children, especially neonates, with suspected CHD. Minor CHD is more difficult to consistently diagnose accurately

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but this does not impact on immediate patient management. It is essential to appreciate the limitations of this diagnostic technique. Hands-on assessment of the patient should not be delayed if major CHD is clinically suspected but cannot be confidently diagnosed or excluded. Telemedicine is utilised more frequently and is more cost effective the greater the distance between DGH and the specialist centre.

MPW-21

Evaluation of the influence of left lateral thoracotomy on scoliosis development in children with aortic coarctation

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Introduction: Surgery in the thoracic area in children has been implicated in the development of scoliosis of thoracogenic origin, particularly lateral thoracotomy is involved in the development of thoracic scoliosis in children with.
The aim of this study was to determine the influence of lateral thoracotomy on scoliosis development in patients with aortic coarctation.
Methods: A group of 64 patients with aortic coarctation was evaluated. 7 subjects were excluded from the study during the examination procedure. 45 patients were operated using left lateral thoracotomy. The mean age for the operative group was 22.7 years (from 9 to 51 years). The age at the time of the operation ranged from 1 month to 16 years (average age 6.9). The mean follow up was 14.8 years (between 7.7 years and 39 years). 12 patients were treated using balloon dilatation and stent implantation. The average age for the non-operative group was 22.9 years (from 11 to 55 years). The mean age at the time of the ballooning and stenting procedure was 9.1 years (between 1.2 and 16.5 years). As a control 17 subjects from the average population were used. A spinal examination together with the evaluation of chest roentgenograms and spinal roentgenograms was conducted.
Results: There was clinical scoliosis in 62% of operated, in 25% of non-operated patients and in 5.8% of the control group. Scoliosis ranged between and . 89% of operated patients with scoliosis had thoracic curves and in 48% of them left sided curves were found. All curves were right sided in non-operated subjects. Scoliosis in the operated group occurred in 63% of males and in 60% of females.
Conclusions:

1. Prevalence of scoliosis after thoracotomy was significantly higher than in the average population and after non-surgical methods of treatment of aortic coarctation.
2. The rate of single thoracic and the rate of left thoracic curves in patients after thoracotomy is higher than the rate in idiopathic scoliosis.
3. The rate of males after thoracotomy with scoliosis is higher than the rate of males with idiopathic scoliosis.

MPW-22

Assessment of left ventricular size and function using 3D-echo-generated volume-time-curves in small infants with severe left ventricular outflow tract obstruction

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Introduction: Precise assessment of left ventricular size and function is of vital importance for the management of severe left ventricular outflow tract obstruction. In borderline cases, reliable echocardiographic measures for biventricular vrs. univentricular repair do not exist. In this setting we assessed 3D-real-time (3D-RT) echocardiography for quantification of left ventricular (LV) function.
Methods: Prospective, longitudinal study of 6 neonates with critical aortic stenosis (AS) and 6 neonates with small left ventricle, hypoplastic aortic valve, aortic arch obstruction and VSD (AAO+VSD), compared with age- and body surface matched healthy controls – using 2D- and 3D-RT-echo (Sonos 7500; Q-Lab 4.2, Philips). Evaluation of: LV-volume, ejection fraction (EF) and 3D-volume-time-curves. Peak ejection and peak filling rate (PER, PFR), time to peak filling and ejection rate (TPFR, TPER) were determined from the 3D-volume-time-curves using a newly developed algorithm VTC 1.5 (Figure) and normalised to body surface and enddiastolic volume.
Results: The median follow up was 12 months [range 4–24]. In neonates with critical AS, initial systolic function was profoundly disturbed with paradoxical segmental movement and reduced EF and PER. The deranged diastolic function was reflected by a reduced PFR and TPFR. After relief of the LV outflow tract obstruction, LV systolic and diastolic function improved gradually, resulting in symmetrical segmental movement and nearly normal EF, PER, PFR and TPFR. In 2 children with persistent reduced diastolic function an univentricular repair had to be performed.
In contrast to critical AS, children with small LV+AAO+VSD presented with almost normal systolic and diastolic function and - after relief of outflow tract obstruction – the LV showed adequate growth compared to normal controls.
Conclusions: In contrast to 2D-echo, 3D-RT-echo allows to determine LV volume irrespectively of ventricular geometry. 3D-generated volume-time-curves give additional quantitative data of systolic and diastolic LV function. Concerning biventricular repair in critical AS, persistent diastolic dysfunction was a negative predictive factor in long term. Prospective studies are needed to confirm these data in a larger cohort and to determine the prognostic value of 3D-echocardiography for therapeutic decisions.

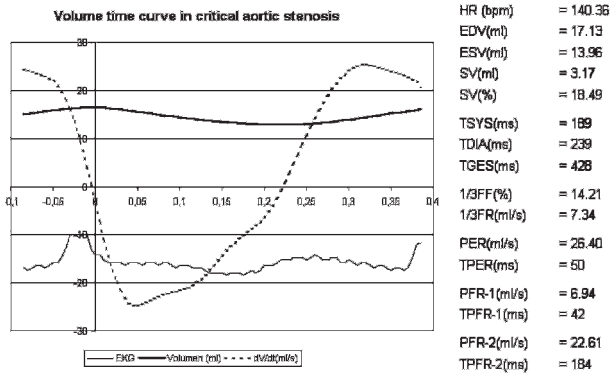

Figure: Volume-time-curve and its 1st derivate in a neonate with critical aortic stenosis

MPW-23

Long term results of early total surgical repair of tetralogy of Fallot

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Objectives: To assess long-term results and outcome in patients (pts) undergoing surgical total repair of tetralogy of Fallot (TOF) at less than 2 years (y) of age with follow-up more than 5y.
Patients and Methods: Total surgical repair of TOF was performed in 385 pts for 11y period 1990–2001 at our institution. Of them 184 (48%) were <2y of age, 111pts (60%) had follow-up >5y (mean 8.7 ± 3y (range 5–15.2) and are included in the study. Only 2pts had preceding palliative systemic-to-pulmonary artery shunt. Transannular patching (TAP) was used in 74pts (63%), patch up to annulus in 33pts (28%) and no patch in 10pts (9%). All pts had clinical assessment, ECG, echocardiography and 55 underwent 24h ECG, 18 – cardiac catheterization, 8–exercise test.
Results: The hospital mortality rate for all pts operated at <2y of age was 3.3% (6pts). There was one late death of systemic infection (0.9%) and no sudden cardiac death. Actuarial survival was 99.1% at 5y and 10y. Early postoperative result was excellent in 94.6% (105pts). Two pts underwent early reoperation because of significant residual VSD and right ventricular outflow tract obstruction (RVOTO). Ten pts (9%) had transitory complete AV block, no patient required early permanent pacemaker (PM) implantation, 1pt required PM for a late complete AV block 10y post operation. In the long term follow-up essential abnormalities are right bundle-branch block (67%), significant ventricular arrhythmias (1.8%) and pulmonary insufficiency (PI) which was mild or moderate in the majority of cases (93%). All pts with severe PI had TAP. No patient with infundibular patch or no patch had severe PI. A late reoperation was performed in 5.4% (6pts) because of significant RVOTO (2 pts) and severe PI (4 pts). "Hybrid" approach was used in 1pt. Two pts required stent implantation.
Conclusions: Early total surgical repair of TOF is confirmed to be safe and effective with low mortality rate and excellent survival. Long-term results are mostly excellent or good. PI is an important determinant of outcome. Infundibular patch or no patch repair do not result in severe PI or RV dysfunction.

MPW-24
Plasma levels of B-type natriuretic peptide at mid- and long-term follow-up after total cavopulmonary connection
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Objective: B-type natriuretic peptide (BNP) is an established diagnostic marker in congestive heart failure in adults and children. In addition, BNP has diagnostic and prognostic value in various congenital and acquired cardiac diseases. The aim of this study was to assess plasma BNP in patients with univentricular heart late after volume unloading by modified Fontan procedure.
Methods: From 2002 to 2007, Plasma BNP concentration was measured by sandwich immunoassay (Triage BNP assay, Biosite®) in 58 patients (34 females) aged 4 to 34 years (median 14.5 years) 0.3 to 22.2 years (9.2 years) after modified Fontan procedure. BNP levels were compared with age and gender-specific normal values, clinical and echocardiographic data, and results of exercise testing.
Results: Plasma BNP had a wide range of 5 to 290 pg/ml, but BNP was normal in 44/58 patients (75%), median BNP was only 13 pg/ml, and interquartile range was 6 to 27 pg/ml. There was no difference between male and female patients, and no correlation was found between plasma BNP and age, postoperative follow-up period, maximum exercise capability, peak oxygen uptake, or blood oxygen saturation during exercise testing. 8/58 patients had BNP levels >50 pg/ml. 2/8 patients had no identifiable problem; protein loosing enteropathy was found in 3/8 patients; significant atrioventricular regurgitation and tachyarrhythmia, heart failure, and cirrhosis of the liver in additional 3/8 patients. 3/8 patients died within 2 years. Longitudinal evaluation of plasma BNP was performed in 14 patients. After 0.6 to 5 years, BNP plasma concentration was at the same level in 11/14 patients and had significantly decreased in one patient. BNP had increased from 13 to 56 pg/ml within 1.8 years in one patient who had developed protein loosing enteropathy, and from 72 to 170 pg/ml within 1.5 years in one patient with decreasing myocardial function.
Conclusion: BNP plasma concentration was normal in the majority of patients up to 18 years after modified Fontan procedure. Elevated and increasing BNP levels were associated with specific sequelae. Measurement of BNP plasma can be useful in the follow-up of patients after total cavopulmonary connection.

MPW-25
Ablation Success and Long Term Outcome in Catheter Ablation of Atrial Tachycardias in Patients after the Fontan Operation
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Introduction: Although catheter ablation in patients after Fontan operation has a quite high acute success rate, data on long-term outcome is limited and recurrence rates up to 30% are reported. We therefore evaluated outcome in our patient population and analysed factors that might influence the risk of recurrence.
Methods: The data of 45 patients (pts) after the Fontan operation (30 male; mean age 24.1 years) with atrial tachycardias were retrospectively reviewed. In the 45 pts, 87 electropyhsiology (EP) studies had been performed. Mean time interval from operation to first EP study was 17.3+/−4.2 years. Complete follow-up was available in 41/45 of pts (91%). Of these, 37 pts were alive and 4 pts (8.8%) had died during the follow-up period. Catheter mapping (using a 3D mapping system) was performed in 104 arrhyhtmias 54% intraatrial reentry tachycardia IART, 33% focal atrial tachycardia FAT, 13% other).
Results: Catheter ablation was acutely successful in 84/104 arrhythmias (82%). Detailed data is shown in Figure 1. Age of the patient, age at operation, number of previous EP studies or hemodynamic parameters did not influence acute success ($p = n.s.$). Ongoing clinical arrhyhthmia at the beginning of the EP study improved the acute success rate significantly (89.2% vs 67.5%, $p < 0.05$).
The mean follow up time since the last EP study is 47 months (+/−21 months). Recurrence rate for the same clinical tachycardia was 26%, whereas the overall recurrence rate of any tachycardia was 50%, developing after a mean of 21 months. Neither age of the patient, age at operation or hemodynamic parameters influenced recurrence rate. Presence of tachycardia at the beginning of the EP study and a focal mechanism lowered recurrence risk, whereas a longer time interval since operation increased recurrence risk ($p = 0.014$).

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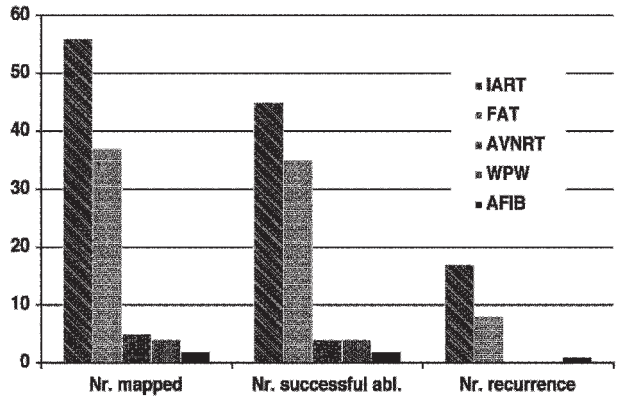

Conclusions: Catheter ablation during ongoing clinical arrhythmia improves both, acute ablation success and freedom of recurrence. Therefore in our opinion conversion of the arrhyhtmia prior to an EP study should be avoided aiming at prompt treatment in the EP lab. As age, prior ablations and the hemodynamic status do not significantly influence outcome, they should not influence the decision about ablation.

MPW-26

Transcatheter closure of congenital ventricular septal defects in a GUCH: mid term results and complications

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Introduction: The safety and efficacy of transcatheter VSD closure in children has been previously reported.
This report outlines mid-term follow-up results and complications in adult patients that underwent transcatheter VSD closure.
Methods: Between January 2000 and June 2006 we prospectively collected data on 145 patients who underwent transcatheter closure of a VSD at our institution. Forty of these 145 subjects were adults. A shunt was considered significant when the following were found: (i) left atrial enlargement, defined as a left atrial-to-aortic ratio >1.5; (ii) left ventricular enlargement (left ventricular overload), defined as a left ventricular end-diastolic diameter >+2 standard deviation (SD) above the mean for the patient's age. Another inclusion criteria was a previous episode of endocarditis. The two different Amplatzer devices available were used. All subjects underwent clinical examination, electrocardiography, chest-X-rays, 24 h EKG-Holter monitoring, and TTE before discharge and at 1, 6, and 12 months after the procedure and yearly thereafter.
Results: 41 procedure were carried out in 40 patients; a mVSD-O was used in 22 pts and a pVSD-O in 18 pts (1 pt had two devices inserted). No deaths occurred; any procedure was aborted.
A total of 6 (14.6%) complications occurred. The most frequent complication was a rhythm abnormality (n = 4). No device embolization occurred. The median duration of follow-up was 36 months (range: 6–81 months). No deaths or cases of endocarditis occurred.
Conclusions: The percutaneous technique of VSD closure is widely appreciated by patients and their parents because it has less psychological impact (given the absence of a skin scar), the time spent in hospital is shorter, the procedure causes less pain and discomfort, and there is no need for admission to an intensive care unit. In adult patients this technique is also better than in young subjects with less complications and with no evidence of cAVB. Post operative residual VSDs can be successfully treated with the benefit of avoiding further surgery.
Greater experience, possibly in a multicenter trials, and long-term follow up are required to better assess the safety and effectiviness of this procedure as an alternative to surgical approaches in adult patients.

MPW-27

Effects of Percutaneous Closure of Secundum Atrial Septal Defect in Adults on Ventricular Morphology and Function Examined by Tissue Doppler Echocardiography.

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Aim: To assess impact of transcatheter closure of ASD II on right and left ventricular morphology and function in adults.
Methods: 46 pts, all in sinus rhythm (age 40.3±14.3), 32 f, with ASD II had successfully implanted Amplatzer device. Comprehensive 2D and Doppler echocardiograms were obtained before the procedure and on day 1 and 1, 3 and 12 months thereafter. End systolic (ES), end diastolic (ED), right (RV) and left ventricular (LV), cavity areas (CA) and fractional area change (FAC) were obtained. Maximal diastolic (E', A') and systolic (S') velocities of lateral annulus of mitral (ma) and tricuspid (ta) valves were measured by TDI.
Results: Results are presented in Fig I and II. Statistically significant reduction of systolic and diastolic velocities of both annuli, RVEDCA, RVESCA with simultaneous increase of LVEDCA in all examined pts were observed. The systolic and diastolic velocities of tricuspid annulus were higher then those of mitral annulus in every study. RV FAC diminished 2 days after the procedure with subsequent increment during follow up. Improvement of LV FAC accompanied the immediate increase of LV end-diastolic cavity area.

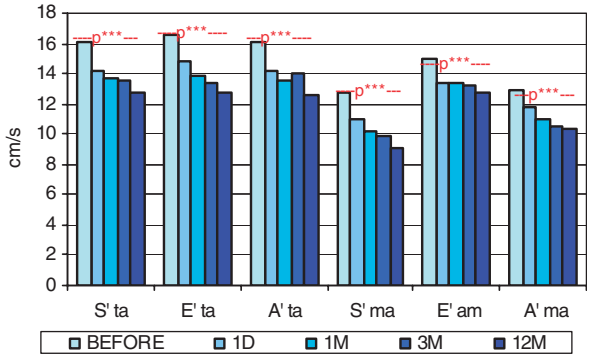

Fig I ***p < 0, 001, **p < 0, 01, *p < 0.05

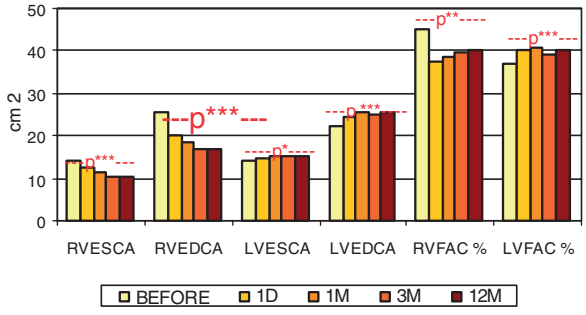

Fig II ***p < 0, 001, **p < 0, 01, *p < 0.05

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Conclusions: Successful percutaneous closure of ASD II resulted in reverse remodelling of both ventricles immediately after the procedure. RV systolic and diastolic areas were decreasing significantly throughout the follow-up. The procedure resulted in gradual, significant decrease of systolic and diastolic velocities of both annuli. Increase of LV function assessed by FAC was accompanied by decrease of MV annulus velocities.

MPW-28

Predictors of ventricular arrhythmia in adult patients with congenital heart diseases.

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Introduction: In adults with congenital hart disease (CHD) ventricular arrhythmia (VA) is not common complication but may lead to sudden cardiac death.
Aim of the study: The assessment of risk factors for VA in adults with CHD during long-term outcome.
Material and method: 1304 patients (P) (586 men) aged 18–72 (mean 29.4 ± 10), were studied and observated during 1–10 years (mean 3.52 ± 1.83) (1995–2004). In P 25 different types of CHD were diagnosed – 13 simple: ASD, VSD, PDA, bicuspid aortic valve, pulmonary stenosis, subvalvular aortic stenosis, VAC, partial anomalous pulmonary venous connection, Marfan's syndrome, mitral insufficiency, idiopathic dilatation of the pulmonary trunk, additional vena cava superior sinistri, Wiliamas syndrome and 12 complex CHD: coarctation of the aorta, Fallot's tetralogy, ASDI, CAVC, Ebstein syndrome, univentricular heart, CCTGA, DTGA, DORV, pulmonary atresia, coronary fistula, Bland White Garland syndrom. The presence of ≥30 monomorphic ventricular beats per hour were considered to be significant. Anatomical complexity of the CHD, performed cardiac operation, hart failure (NYHA > I), cyanosis, age and gender were assessed during first visit. Kaplan- Meier curves were estimated, log-rank tests to compare curves, and Cox proportional hazards model for assessing multivariate associations between VA and risk factors were calculated.
Results: VA was observed in 6.1% of P. The probability of VA occurence after 2 and 5 years was 2.3% and 10.2% respectively. In univariate analysis the probability of VA presence was higher in patients: with complex CHD as compared to simple CHD ($p = 0.00001$), with NYHA > I than in NYHA I ($p = 0.00001$), and in those with cyanosis ($p = 0.00001$). Previous cardiac operation and gender did not have the influence. The most predictive VA risk factor was anatomical complexity (HR = 2.55). Other factors did not rich statistical significance although cyanosis and NYHA > I increase the risk for analyzed arrhythmia occurrence in the whole populations as well as in the subgroups with simple and complex disorders.
Conclusions: Anatomical complexity is a significant risk factor for ventricular arrhythmia occurrence in adults with CHD. Cyanosis and heart failure increase the risk of its occurrence. Previous cardiac operation, age and gender did not have the influence.

MPW-29

Social Aspects of the Life of Adults with Congenital Heart Defects

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Introduction: Due to medical advances we are now about to reach the point where the number of adults with congenital heart disease (CHD) will exceed the number of children with CHD. To date, data on the social achievements of adults with CHD are scanty. Thus, there is an urgent need for information on specific social challenges experienced by this growing population.
Methods: We sent a postal questionnaire to 2,605 adult patients of the National Register for Congenital Heart Disease. With a response rate of 65% the participation was high. The median age was 28 with a range from 18 to 86 years.
Results: 49% had a surgical intervention, 66% of them underwent a re-intervention. 53% are cared for by paediatric cardiologists. 59% live in a partnership, 32% are married and 33% have children. 28% are high school graduates and 11% have a university diploma. Both rates are statistically higher than in the general population. 53% are working full-time, which is significantly less than the general population (57%). Nevertheless, the rate of unemployment is not significantly increased (11% vs. 9%). This is due an increased rate of part-time work (18%) and of early retirement (14%). However, 25% of all adults felt restricted in the choice of their profession, this number is rising to 53% in the group of the complex defects. 26% were denied the possibility of a private health care and 35% were excluded from life insurance from private providers.
Conclusion: The social integration into society is an important component of the quality of life of our patients. Therefore, the goal of medical care is not only to achieve long-term survival but also to achieve the best possible social outcome. In our big and nationwide sample we found a high academic performance, but only moderate employment rates. Therefore, patients and in particular those with more complex forms would benefit from an individual and specific career counselling. The wish of adults with CHD for financial security through private insurance providers has to be considered and discussed.

MPW-30

Quality of life and exercise capacity in 497 patients with congenital heart disease

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Objective: to compare self reported health related quality of life with objective exercise performance in patients with congenital heart disease.
Patients and methods: 497 patients (219 female, 14–73 years old) with various congenital heart defects (52 shunt, 62 left heart obstruction, 29 right heart obstruction, 42 Ebstein, 86 Fallot, 84 TGA after atrial switch, 40 other TGA, 30 Fontan, 18 palliated/native cyanotic, 54 others) as well as a group of 52 healthy controls (18 females, 14–57 years old) completed a health related quality of life questionnaire (SF-36). Then they performed a symptom limited cardiopulmonary exercise test with measurement of oxygen uptake on a bicycle in a sitting position. As patient groups differed in sex and age all measured values were normalized to published sex and age related reference values.
Results: Despite severe limitations at the exercise test, there was only reduced quality of life in the scales of physical functioning ($p < 0.0005$) and general health ($p < 0.0005$). This could be confirmed in all diagnosis subgroups. There was very good correlation of self estimated physical function ($r = 0.402$, $p < 0.0005$), physical role ($r = 0.157$, $p < 0.0005$), general health ($r = 0.302$, $p < 0.0005$), vitality ($r = 0.173$, $p < 0.0005$) and social functioning ($r = 0.159$, $p < 0.0005$) with peak oxygen uptake.

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Bodily pain, emotional role, mental health and health transition showed no significant relation.
Conclusion: Patients with congenital heart defects are only impaired in their physical functioning and their general health in respect to their quality of life. This is also true for severe heart defects such as Fontan or palliated cyanotic defects. The amount of impairment correlates directly to the exercise performance.

MPW-31
Ventricular repolarisation versus transvalvular gradient and left ventricular mass in children with aortic valve stenosis
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Congenital aortic valve stenosis (AS) could lead to life threatening arrhythmia. Abnormalities of the ventricular repolarisation participate in the mechanism of the severe arrhythmia.
Aim: The aim of this study was to analyze dyspersion of QT interval (QTd) in children with different stage of AS and with different left ventricular mass.
Material and methods: 60 children with AS aged 5–18 years (AS group) and 60 matched healthy children (Control group) participated in this study. AS group was divided into 3 subgroups according to transvalvular gradient (PG) obtained in ECHO+Doppler: subgroup I – 21 children with PG < 40 mmHg, II – 27children with PG 40–69 mmHg, III – 12 children with PG >70 mmHg. QTd was calculated in standard 12 leads ECG. Left ventricular mass was calculated according to the Deveroux formula in ECHO and indexed by body surface area (LVMI). Holter ECG monitoring was performed for arrhythmia evaluation. Results in AS group were compared with Control and in subgroups I, II, III between each other. Corelation coefficients (r) between QTd and PG, QTd and LVMI were estimated.
Results: Values of QTd in AS group, Control and subgropus I, II, III are presented in table.1

Table 1.

	AS group	Control group	p	I subgroup	II subgroup	III subgroup	p
QTd (ms)	44 ± 14	29 ± 7	<0,0001	38 ± 12	43 ± 15	55 ± 10	I vs III: < 0,005 II vs III: < 0,01

The stronge corelation between LVMI and PG ($r = 0.64$ $p < 0.001$) was found.
LVH was diagnosed in 17 children (3 children from subgroup I, 5 patients from subgroup II, 9 children from subgroup III).
Corelations between QTd and PG and between QTd and LVMI are presented in table 2

Table 2.

	QTd	
Parameter	r	p
PG	0.42	<0.001
LVMI	0.50	<0.0001

Ventricular arrhythmia was diagnosed in 20 children with AS: 4 children from subgroup I, 7 patients from subgroup II and 9 children from subgroup III.
Conclusions
1. Dyspersion of QT is higher in children with AS than in healthy children and increases with the growth of transvalvular gradient and left ventricular mass.
2. Increased dispersion of QT in children with aortic valve stenosis may reflect higher risk of arrhytmia.

MPW-32
Transcather closure of congenital and acquired ventricular septal defects with Amplatzer devices
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Introduction: Transcatheter closure of congenital and acquired ventricular septal defect (VSD) with Amplatzer occluders has been recently introduced as an alternative to surger.
Material and methods: Transcatheter closure of VSD was attempted in 45 patients (pts) aged from 0.8 to 81 y. There were 17 pts with perimembranous VSD (PmVSD), 7 with muscular VSD (MVSD), 20 with postinfarction VSD(PIVSD) and 1 pt with posttraumatic muscular VSD. Following Amplatzer devices were used: asymmetric (PmVSDO), muscular (MVSDO), Atrial septal occluder (ASDO) and postinfarct VSD occluder (PIVSDO).
Results: The procedure was succesfully finished (without residual shunt) in 15 out of 17 pts with PmVSD. In 7 – PmVSDO and in 9 – MVSDO was applied. In 2 pts the procedure was abandoned because of rhythm disturbances (during introduction of delivery system in one and just after PmVSDO placement in the second).
In 4 out of 7 pts with muscular VSD after MVSO implantation small residual leak through an additional VSD persisted. Early embolization of MVSDO occurred in 2 pts with MVSD. One of them had complex CHD (subsequent surgery), another – 40y old male with coexistent arterial hypertension had a thick muscular septum. Early embolization od MVSDO also occurred in a young male with posttraumatic muscular VSD. The IVS was also in this case thicker than the waist of the device (MVSDO). In both pts the devices were retrieved from the aorta with a bioptom and a lasso respectively.
Out of 20 pts with PIVSD in 15 ASDO were applied (in 2 pts – 2 ASDO), in one – MVSDO, in 2 – PIVSDO and in one cribriformis ASDO. The procedure was abandoned in 3 of them because of technical problems. One pt died during the procedure because of acute tamponade.
Conclusion: Transcatheter closure of congenital and acquired VSD is still a difficult but promising procedure and in some patients can be an alternative to surgical treatment. Technical strategy and choice of optimal device should be thoroughly studied in every case, since ideal devices have probably not been invented yet. Muscular VSDO device is not an optimal one for transcatheter closure of muscular VSD in adults.

MPW-33
Repeat percutaneous pulmonary valve implantation: proof of principle as long-term management strategy for RVOT conduit dysfunction
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Introduction: The longevity of surgically placed conduits for RVOT dysfunction can now be prolonged through percutaneous pulmonary valve implantation (PPVI). However, the longevity of PPV devices will also be limited with time. The treatment of failing PPVI with repeat PPVI offers the potential to prolong the

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intervals between surgeries needed or even avoid surgery entirely for patients who typically require multiple re-operations.
Methods: We retrospectively reviewed the 15 patients who underwent repeat PPVI between September 2000 and October 2006 as treatment for early device failure ('Hammock effect' [5], stent fracture [8], residual stenosis [2]). All patients were assessed before and after PPVI according to a detailed protocol that included clinical assessment, antero-posterior and lateral chest X-ray and trans-thoracic echocardiography (VIVID 7, GE, Medical Systems, Milwaukee, Wis.). Patients with residual or recurrent RVOT dysfunction were identified and their subsequent management and follow-up documented.
Results: Repeat PPVI was feasible in all patients and there were no procedural complications. RVOT gradient and right ventricular systolic pressure (RVSP) could be reduced effectively (RVOT: 48.8 ± 5.0 to 17.6 ± 2.2 mmHg, $P < 0.001$ and RVSP: 75.7 ± 6.0 to 46.3 ± 3.2 mmHg, $P < 0.001$). Moreover, repeat PPVI achieved better lowering of RVSP compared to initial PPVI (−29.4 ± 3.8 vs. −12.8 ± 4.9 mmHg, $P = 0.01$). During follow-up, 3/15 required re-intervention (third PPVI: stent fracture [n = 1], device explantation: external compression by the sternum [n = 1], endocarditis [n = 1]). In the remainder, second PPVI resulted in a sustained improvement in hemodynamic and functional status with a mean follow-up of 8.3 ± 2.1 months.
Conclusion: Repeat PPVI is feasible, safe and effective treatment for early device failure and provides proof of principle as potential long-term management strategy for RVOT dysfunction.

MPW-34
Transesophageal echocardiography in newborns with miniaturized transducer to guide interventional cardiac catheterization
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Introduction or Basis or Objectives: The role of transesophageal echocardiography (TEE) to guide interventional cardiac catheterization is well documented. It has limitations in newborns and infants with hemodynamic or respiratory compromise. We report our experience with the Siemens Acunav probe in the performance of TEE in newborns and infants in these type of procedures.
Methods: Retrospective review of all TEE performed between Febrery 2005 and January 2007 with the Acunav Siemens probe, 10f (3.2 mm diameter), monoplane, with Doppler and colour capabilities, to guide interventional cardiac catheterization in newborns and infants.
Results: 22 studies were performed in the inclusion period. Median age was 38 days (range between 0–139 d). Median weight was 3609 gr (range between 1800–6400 gr). All the patients were under mechanical ventilation and 4 in ECMO. 20 cases were performed in the catheterization room (90.9%) and 2 in the Neonatal Intensive Care Unit (9%). The procedure was: 1) Creation of an atrial septal defect in 12 cases (54.5%): 7 radiofrequency catheter perforation and stent implantations, 2 cutting ballon and posterior ballon dilatation and 3 Rashkind atrial septostomy. 2) Pulmonary Ballon valvuloplasty in 6 cases (27.2%) and 3) Aortic ballon valvuloplasty in 4 cases (18.1%).
The procedure was well tolerated and there were no complications related to the probe implantation. The images were of goog quality in 100% of the cases although in one newborn with Transposition of the great arteries they were worse than transthoracic echocardiography for an atrial septostomy.
Conclusions: This miniaturized transducer designed initially for intravascular use is safe and useful for performing TEE to guide in interventional cardiac catheterization in newborns and infants with hemodynamic or respiratory compromise. It has a limitation in the evaluation of ventricular septum and auriculo-ventricular valves but this is not necessary in these procedures.

MPW-35
Evaluation of residual shunt at mid-long term follow-up of percutaneous closure of patent foramen ovale
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Aim: Evaluation of residual right-to-left shunt (RS) at mid-long term follow-up (FU) after transcatheter occlusion of patent foramen ovale (PFO).
Patients and method: Study population consists of 198 patients (mean age: 42 ± 19) with percutaneous treatment of PFO (104 pts) and PFO associated with fenestrated atrial septal aneurysm (FASA) (94 pts) with FU period at least of one year.
Our protocol of FU of PFO closure consists of contrast-enhanced transthoracic echocardiography (ce-TTE) at discharge, 6 months and one year after the procedure. In case of RS at 1 year, patients are evaluated with contrast-enhanced transcranial Doppler (ce-TCD) which is repeated every 6 months in case of persistent RS. The patients with large RS (DTC > 30 microbubbles) at 2 years FU are investigated with transesofageal contrastography (ce-TEE) and x-ray angiography.
Results: At one year FU, RS was detected with ce-TTE in 14 asymptomatic patients (7%), 12/104 FASA (12.5%) and 2/94 PFO (2%), $p = 0.001$. In 13 patients RS was confirmed and in 1 excluded by ce-TCD. At 2 years FU, in 11 RS persisted, large in 8 (7 with ASA and 1 with PFO) and trivial in 3; in 2 cases RS disappeared. In 5/8 patients with large RS (4 ASA and 1 PFO), ce-TEE and angiography were performed: in the patient with PFO no RS was detected; in 1 patient an accessory hole was found and successfully closed with another device; in 2 patients, with multifenestrated ASA and Amplatzer cribriform device, a trivial RS across the PFO was found and left untreated; in 1 patient no accessory fenestrations was detected and the trivial RS was related to incomplete device endothelization. Three patients with large RS are waiting for the evaluation.
Conclusions: Significant RS after transcatheter closure of PFO is rare and complex anatomy of the atrial septum is a risk factor. An accurate evaluation of presence and degree of RS is mandatory and ce-TCD has high sensitivity and low invasivity and is probably the method of choice. In case of significant RS transcatheter closure of residual fenestration can be possible.

MPW-36
Interventional closure of fenestration as a final stage of treatment in HLHS patients after Fontan operation
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Background: Creating of fenestration between the tunnel of inferior vena cava and right atrium cavity leads to significant decrease of postoperative failures in Fontan operation. The fenestration reduces the risk of peritoneal transsudate in patients with univentricular anatomy treated by Fontan operation. After full adaptation to Fontan circulation in this patients (pts) interventional closure of fenestration using the septal occluders can be performed.
Aim: We estimated the efficiency of interventional fenestration closure in HLHS pts after Fontan procedure, and right ventricle (RV) function after intervention.
Material and Methods: Between July 2004 and December 2005 we interventionally closed fenestration in 10 pts with HLHS after Fontan procedure. We assessed 8 pts (6 males, 2 females) using physical examination, saturation monitoring, echocardiographic and angiographic evaluation. The extracardiac Fontan procedure as an end-stage of HLHS treatment was performed in children aged from 20 to 68.5 months (mean 47 months). The transcatheter fenestration closure was performed 9.5 to 36 months (mean 21 months) after Fontan procedure. Cyanosis and hypoxemia was noted prior to procedure. RV dimension, tricuspid regurgitation (mild in 4 pts, moderate in 4 pts) and RV function assessed by RV myocardial performance index (RV-MPI)) were determined in echocardiographic examination. Fenestration closure was performed using Amplatzer occluder (diameter: 4 mm and 5 mm) in 7 pts and Star-Flex device number 17 in 1 pt.
Results: We observed cyanosis reduction and significant mean oxygen saturation increase from mean 78% (74–83%) to mean 94% (91–98%) ($p < 0.0001$) after fenestration closure. No significant changes in RV dimensions or tricuspid regurgitation were noted in echocardiographic examination after intervention, but increase of RV-MPI from mean 0.406 (0.247–0.642) to 0.553 (0.254–0.723); $p = 0.04$) was observed. Insignificant mean central venous pressure increase from 1 to 2 mmHg (in 1 case −5 mmHg) occured.
Conclusions: Interventional fenestration closure in HLHS patients after Fontan operation is a safe and efficient method. Fenestration closure leads to arterial blood saturation increase with no significant venous pressure increase in direct observation. RV-MPI significantly increases after fenestration closure. This index is additional parameter useful in RV function assessment.

MPW-37

Transposition of the great arteries and left ventricular outflow tract obstructions – is there a place for switch operation?

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Objective: To determine, through hemodynamic and angiocardiographic study, the types of left ventricular outflow tract obstruction in patients with transposition of great arteries.
Patients and Methods: Two hundred and forty six patients with transposition of great arteries were studied in three institutions in Rio de Janeiro – Brazil. In all of them, the contrast injection was done in left ventricle in axial projection because subtle forms of left ventricular outflow tract obstruction can be missed with conventional angiocardiographic views, since the anterior wall of the left ventricle may obscure the left ventricular outflow tract obstruction. In addition, the type and size of ventricular septal defect, the size of pulmonary annulus in diastole and left ventricular-pulmonary artery gradient were analyzed.
Results: In 27 out of 246 patients the left ventricular outflow tract obstruction type was: G1: sub-pulmonary fibro-muscular tunnel – 10 patients (37%); G2: pulmonary valvular stenosis – 4 patients (14.8%); G3: dynamic sub-pulmonary stenosis – 5 patients (18.5%) and G4: sub pulmonary fibro-muscular ring – 8 patients (29.7%). All patients out G3 and one in group G2 had mal-alignment ventricular septal defect. Left ventricle systolic pressure was at systemic level in patients groups G1, G2 and G4. The pulmonary valvular annulus was expressively larger in groups G1 and G4 then in groups G2 and G3.
Conclusions: Precise angiocardiographic assessment of the left ventricular outflow tract obstruction in transposition of great arteries is essential since the surgical technique depends on the type and severity of left ventricular outflow tract obstruction.

MPW-38

Long term evaluation of coronary arteries in patients after the anatomical correction of transposition of the great arteries

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Introduction: Anatomic correction is nowadays a method of choice for treatment of transposition of the great arteries (TGA). A crucial point of this operation is relocation of the coronary arteries in such a way so that they do not twist or kink, allowing proper perfusion to the heart muscle.
Methods: Between 1992 and 2006, 440 patients with TGA were treated with arterial switch operation. In 130 patients, who had undergone ASO in neonatal period, 5 to 8 years following the operation selective coronary angiography was performed.
The aim of the study was to determine the number of patients with significant stenosis of a coronary artery and its correlation with the operation.
Results: In 89 patients (68%) coronary anatomy was typical and without any stenosis.
In the remaining 41 patients mostly different congenital dislocations of coronary arteries were observed. In 5 patients significant stenosis of a coronary artery was detected. In 3 cases a coronary artery was narrow in its distal part which was considered a congenital anomaly.
In 2 cases coronary stenosis was proximal and it was most probably due to the performed operation.
In the first case there was a 30% stenosis of orifice of the left coronary artery.
In the second aortic orifice of the left coronary artery could not be detected. The flow was retrograde from the right coronary artery via the collateral vessels.
In all of the cases ECG was correct.
Conclusions:

1. In most cases angiography performed 5–8 years following ASO shows correct anatomy, without significant stenosis, of the coronary arteries.
2. Of 130 examined patients only two had significant postoperative stenosis of a coronary artery.
3. Late complications of surgical coronary translocation in patients with TGA are rare and can be diagnosed only after performing coronary artery angiography.

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MPW-39

Pre- and postcardiac surgery weight gain of infants under 1 year of age in relation to neurodevelopmental outcome

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Objective: To determine predictors of pre- and postoperative weight gain in infants undergoing open heart surgery in relation to neurodevelopmental outcome.
Methods: In a prospective cohort study pre- and postoperative weight gain was evaluated in infants under 1 year of age undergoing open heart surgery. Body weight was determined at birth, at operation and at one year of age. A standard neurological examination was performed before surgery and at one year of age. At this time children were also assessed with the Bayley Scales of Infant Development II. In a multiple regression analysis, postoperative weight gain and the neurodevelopmental outcome at one year of age were related to preoperative medical, neurological, perioperative and surgery-related factors.
Results: The study cohort comprised 107 infants (54 females) with a median age of 3.9 months (range: 0.1–10.2) at the first cardiac operation. Follow up time was 8.7 months (3.5–15.7). Mean weight at birth was 3.0 ± 0.7 kg, at operation 4.8 ± 1.5 kg, and at follow up 8.7 ± 1.4 kg. The weight gain was preoperative 13.2 ± 9.3 g/d and postoperative 14.3 ± 4.6 g/d, respectively.
Better postoperative weight gain was independently related to lower RACHS-1 score ($p < 0.05$), and to an older age at operation ($p < 0.05$). The neurological outcome at one year of age was influenced by the preoperative neurological findings ($p < 0.001$). Longer hospitalisation time predicted a poorer cognitive performance at one year of age ($p < 0.05$), whereas pre- and postoperative weight gain did not. Poor postoperative weight gain was associated with impaired motor development at one year of age ($p < 0.05$).
Conclusions: In infants, postoperative weight gain after cardiac surgery is mostly influenced by cardiac factors such as type of congenital heart defect and age at operation, whereas the preoperative neurological status and other pre- and postoperative factors of intensive care management are not significant risk factors for postoperative weight gain.
Neurodevelopmental outcome at 1 year is strongly determined by the preoperative neurological status. An adequate postoperative weight gain predicts a better psychomotor development. Shorter hospital stays correlate with better mental outcome.

MPW-40

Dexmedetomidine, a Novel Agent for the Acute Treatment of Supraventricular Tachyarrhythmias after Pediatric Cardiac Surgery

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Background/Objective: Supraventricular tachyarrhythmias (SVTs) can be a cause of increased morbidity and mortality after congenital cardiac surgery. Dexmedetomidine, a primarily sedative agent, has potential novel anti-arrhythmic effects through the central alpha-2 adrenoceptors and imidazoline I2 receptors. We describe our preliminary experience using dexmedetomidine in the acute management of postoperative junctional ectopic tachycardia (JET), ectopic atrial tachycardia (EAT) and AV node reentry tachycardia (AVNRT).
Methods: Data from 8 children that received dexmedetomidine for SVT after cardiac surgery were evaluated. Treatment was considered primary if dexmedetomidine was used as a first line agent or rescue if other anti-arrhythmics were used prior. Patients were followed for 48 hours.
Results: Table 1 shows the baseline and after dexmedetomidine treatment data. Rhythm and rate control was achieved in all patients. Patient one had several breakthrough episodes of hemodynamically significant EAT and received amiodarone bolus 5 mg/kg, with 5–15 mcg/kg/min maintenance for 32 hours without improvement. Eighty-five mins after dexmedetomidine he was in sinus rhythm (SR). Patient 2 had JET that did not improve after 4 hours of hypothermia and 5 mg/kg amiodarone. Two hours after dexmedetomidine, HR decreased to 172 from 184 and within 3.5 hrs to 146. Patient 3, after 35 hrs of controlled rhythm and rate with dexmedetomidine, developed sepsis, multiorgan failure and recurrence of JET. Patients 4 and 6 developed accelerated junctional rhythm (AJR), 166 and 150 b/m respectively. Given the high risk of progression to JET, dexmedetomidine was started and both remained in AJR and overdrive paced. Patient 5 had controlled rate and conversion to SR, but 3 hours after dexmedetomidine was discontinued, AJR and JET recurred. Patients 7 and 8 had resolution of AVNRT within 2 and 30 mins respectively. Core temperature was reduced in all patients to 35 to 36. Four patients received a bolus of dexmedetomidine, 0.5–2 mcg/kg and well tolera ted. The average dexmedetomidine dose, 0.95 mcg/kg/hr, was higher than our previously published data for sedation.
Conclusion: Our data suggest that dexmedetomidine may be an alternative adjunct for the acute management of supraventricular tachyarrhythmias after pediatric cardiac surgery.

Table 1.

	Age-days	Diagnosis	Arrhythmia (HR)	Primary vs. Rescue	Time to HR control (HR#170)†	Time to SR†
1	5	TGA-IVS	EAT(183)	Rescue	0.58	1.4
2	6	TGA-IVS	JET(230)	Rescue	3.5	48
3	38	SV-TAPVRO	JET(170)	Primary	0.5	Remained in AJR
4	125	DORV-PS	AJR(166)	Primary	NA	Remained in AJR
5	7	TA II	JET(178)	Primary	1.5	13
6	87	CAVSD	AJR(150)	Primary	NA	Remained in AJR
7	4	TGA-VSD	AVNRT(214)	Primary	2mins	2mins
8	1	HLHS-PAPV-RO	AVNRT(272)	Primary	0.5	Remained in AJR

	Reccurence of Arrhythmia	Dex. Bolus (mcg/kg)	Dex. Infusion (mcg/kg/hr)	HR	SBP
1	N	0.5	1(0.5–1.25)	124 ± 8	73 ± 8
2	N	2	1(0.4–1.5)	141 ± 18	68 ± 7
3	Y (35hrs)‡	1	1.23(0.4–1.5)	158 ± 9	66 ± 8
4	N	NA	0.9(0.7–1.2)	132 ± 10	87 ± 9
5	N	NA	1(0.5–1.0)	154 ± 5	77 ± 9
6	N	NA	1.22(0.7–1.25)	133 ± 11	78 ± 9
7	N	1 #	0.76(0.5–1)	154 ± 16	65 ± 6
8	N	NA	0.48(0.4–0.7)	159 ± 7	52 ± 4

†:In hours
‡:Developed septic shock.
#:Bolus given over 3 mins

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P-1

Cryoablation versus Radiofrequency Ablation for AV Nodal Reentrant Tachycardia in Children and Adolescents

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Introduction: Atrioventricular (AV) nodal reentrant tachycardia (AVNRT) is the second commonest type of supraventricular tachycardia in children. Transcatheter ablation using radiofrequency current (RFC), is very successful but carries a risk for damage to the AV node. Cryoablation (CR) is a new and potentially safer method of treatment. We sought to compare the two methods in our center.
Methods: Retrospective review of medical records of 20 patients [12 females, 7–18 (13.25 ± 2.59) years old] who underwent RFC (group A) and 20 pts [11 females, 8.5–18 (12.17 ± 3.07) years old] who underwent CR ablation (group B) in our center. Slow pathway was targeted in both groups.
Results: All patients in both groups had typical AVNRT. Three pts in group B had additional arrhythmogenic substrates (ectopic atrial tachycardia, nodoventricular fiber and accessory pathway). Procedure duration was 90–210 min (146.05 ± 37.36) in group A and 120–480 min (184.40 ± 75.78) in group B ($p<0.05$). Number of lesions was 1–26 (8.85 ± 6.63) in group A and 1–10 (3.95 ± 2.39) in group B ($p<0.05$). Maximum temperature was 47–62°C (54.80 ± 3.88) in group A and minimum temperature was −70 to −85°C (−79.85 ± 4.35) in group B. Fluoroscopy time was 2.3–27 min (14.33 ± 8.2) in group A and 0.5–31.2 min (6.41 ± 6.91) in group B ($p<0.05$). The NavX system was used in all group B pts and in 10 group A pts. Success was achieved in 20/20 pts in group A and in 19/20 group B pts ($p = 0.14$). One group B pt required left-sided slow pathway ablation with RF, and one had transient AV block with CR and the procedure was terminated. There was 1 pt with transient AV block in group 1 and 2 pts with transient AV block in group B, with no other complications. Follow-up was 1–4 yrs for group A and 2 mo–1.5 yrs for group B. Recurrence rate was 2/20 (10%) in group A and 2/19 (10.5%) in group B ($p = 0.34$).
Conclusions: Cryoablation offers comparable results to RFC ablation for treatment of AVNRT in children. Because of the reversible effect of CR during the initial phase of cryoenergy application, the risk of damage to the AV node appears to be very low.

P-2

Long-term experiences of AutoCapture controlled pacing in children

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Introduction: Automatic threshold tracking with adjustment of stimulation output provides extended battery service life in adults. Experiences with epicardial leads in paediatric patients are scarce. Aim of this study was to examine the feasibility of the AutoCapture (AC) algorithm with epicardial leads, and to compare the effects of adjusted output on longevity between AC versus conventionally programmed devices in children.
Patients and Methods: A total of 62 children were prospectively enrolled. In 56 (90%) children (age 8.0 years (0.0–18.4)) devices were AC programmed (AC group) and followed up to 9 years (median 3.0). Six patients with conventionally programming served as a control group. All devices were connected to bipolar steroid-eluting epicardial pacing leads (Medtronic CapSure Epi 10366 or 4968). Data are given as median.
Results: Ventricular thresholds did not differ between the AC and control group (0.70 versus 0.88 Volt@0.5ms, $p = 0.07$). An appropriate ratio between evoked response sensitivity and polarization signal (lead polarization safety margin) at implantation was obtained in the AC group (5.27 (1.7–24.9)). A decrease below 1.7:1 necessitated a switch to conventional settings in 3 children. In the remaining 53/62 (85%), no differences were observed for lead polarization safety margins at first and 4-year follow up (6.9 versus 4.1, $p = 0.07$) and right or left ventricular pacing sites (3.7 versus 5.8, $p = 0.31$). Calculated battery lifetime for single and dual chamber devices with a capacity of 0.95 Ah was higher for the AC than control group (22.2 versus 4.6 years and 13.2 versus 6.4 years, $p<0.001$).
Conclusions: AC controlled pacing is feasible in children with epicardial leads during up to 9 years follow up. A sufficient lead polarization safety margin could be maintained in 85%, independently of the pacing site. Substantial prolongation of the calculated battery service life was observed compared to conventionally programmed devices.

P-3

Heart rate variability and baroreflex sensitivity are related to fat amount in healthy children

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Introduction: Overweight and obese adults have sings of increased sympathetic activity. It is supposed that there is an interaction between fat mass and autonomic nervous system activity. However, this interaction has not been evaluated in children.
Aim: To assess the relation between the amount of fat mass and heart rate variability (HRV) as well as baroreflex sensitivity (BRS) in healthy children.
Methods: 33 healthy children (17 girls; 11–18 years old) were included. The relative (as % of whole body mass) amount of the fat mass was measured by whole-body impedance (Bodystat 1500, USA). To assess the autonomic regulation of the cardiovascular system 30-minute resting ECG (Porti 5, TMSI, Netherlands) and blood pressure (Portapres 2, FMS, Netherlands) signals were recorded in supine position. The HRV was evaluated by spectral analysis (total power of the HRV spectrum (TP), very low frequency (VLF), low frequency (LF), and high frequency (HF) and LF/HF ratio). The BRS was measured using a cross-correlation method. The associations between fat amount and HRV or BRS were calculated with the use of nonparametric Spearman correlation. Statistically significant value was $p<0.05$.
Results: There was a significant negative correlation between the fat amount and mean duration of cardiac cycles ($r = -0.55$; $p = 0.0008$), TP ($r = -0.48$; $p = 0.0046$), VLF ($r = -0.54$; $p = 0.0012$) and BRS ($r = -0.42$; $p = 0.015$). There was no significant correlation between body fat indices and LF, HF as well as LF/HF.
Conclusions: These results suggest that there is a significant link between fat mass and autonomic modulation of cardiovascular system in children. The higher body fat amount is associated with

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a reduced HRV and BRS as well as an increased resting heart rate. It also confirms that sympathetic nervous system activity is increased and/or parasympathetic tone decreased in children with higher fat mass. Further studies are necessary to explain the clinical meaning of this finding.

P-4

Morphologic and Functional Characteristics of the Cited2 Knockout Mouse Embryo: A Preliminary Study

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Introduction: Cited2 knockout murine embryos are known to manifest neural tube and cardiac defects and appear to die at around embryonic day (ED) 15.5. Embryonic death may be associated with deteriorating cardiac function. We aimed to study the morphology and cardiac function of Cited2 mouse embryos and correlate echocardiographic and autopsy findings with genotype.
Hypothesis: We hypothesized that it is possible to identify Cited2−/− murine embryos using ultrasonography allowing longitudinal evaluation of their cardiovascular function in utero.
Methods: Four pregnant Cited2+/− mice (pregnancy resulting from a cross of +/− female and +/− male) were scanned using a Philips Sonos 5500 ultrasound system with a 12MHz probe at ED13.5, 14.5, 15.5 and 18.5, respectively under isoflurane anesthesia. The position of each embryo was mapped in the maternal abdomen. Doppler waveforms were obtained from the cardiac inflow and outflow tracts, descending aorta, ductus venosus and umbilical cord of each embryo. Each dam was sacrificed after the examination. Embryos were isolated and their crown-rump length (CRL), body weight and placental weight were determined. Tail snips were used for genotyping. Embryos were embedded in paraffin, sectioned, stained and histopathological examination was performed.
Results: Twenty-one out of 24 live embryos were evaluated with echocardiography. Eight of them had abnormal echo Doppler-arrhythmia, abnormal outflow tract blood velocities, evidence of atrio-ventricular and semilunar valve regurgitation, prolonged isovolumic contraction time, and/or monophasic ventricular filling pattern. Genotyping showed that 6 embryos were Cited2+/+, 9+/−, and 9−/−. All Cited2−/− embryos had shorter CRL, smaller placentas and weighed less in comparison to their littermates. Eight Cited2−/− embryos that were examined by ultrasonography (one was missed) had abnormal echo Doppler. However, on gross examination, only 3 of them had structural abnormalities (exencephaly, orofacial cleft). Histopathological examination of two Cited2−/− embryos showed evidence of right isomerism.
Conclusion: Cited2−/− mouse embryos have right isomerism and abnormal cardiac function. It is possible to identify accurately the phenotype of Cited2−/− embryos non-invasively with ultrasound which allows longitudinal monitoring of cardiac function in utero providing a marker of congenital heart disease.

P-5

A medium-chain triglyceride diet improves survival under stress but does not improve ventricular arrhythmias in mice with a fatty acid oxidation disorder

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Background: Disorders in long-chain fatty acid metabolism can present shortly after birth with multiple organ dysfunction including cardiomyopathy, arrhythmias, liver dysfunction, and premature death. Children with disorders in fatty acid oxidation are fed diets rich in medium-chain triglycerides (MCT) to prevent organ dysfunction. We used a mouse model of very long-chain acyl-CoA dehydrogenase deficiency (VLCADD) to test responses to this MCT diet on organ function, ventricular arrhythmias and survival during stress.
Methods: VLCADD and Wild Type (WT) mice were fed either a MCT diet or a control diet for 4–8 weeks. Mice were subjected to fasting in the cold (F+C), a combined stress known to induce premature death in VLCADD animals. Tissues were harvested after 4-hours of stress to measure respiration and uncoupling in heart, skeletal muscle, and brown adipose tissue (BAT). Gene expression analysis was performed with real time RT-PCR. Additional mice underwent electrophysiology studies with isoproterenol to assess the effect of diet on ventricular arrhythmias.
Results: Survival and temperature control improved after 4-weeks of MCT diet in VLCADD mice (survival 0% vs. 38% before and after 4-weeks MCT diet). Survival further improved after 8-weeks of diet with survival of 56% of the VLCADD animals. Before MCT diet, VLCADD mice had impaired uncoupling during F+C stress in BAT, but respiration and uncoupling were only mildly affected in heart and skeletal muscle. After MCT diet, uncoupling did not change in BAT of VLCADD mice, indicating that the improvement was due to shivering thermogenesis rather than non-shivering thermogenesis. However, ventricular arrhythmias on isoproterenol were increased after 8 weeks of MCT diet (12 ± 10 vs. 4 ± 3 induced episodes, $P = .013$), whereas ventricular refactoriness (VERP) was reduced after MCT diet (32 ± 10 vs 44 ± 8 msec, $p = 0.026$)
Conclusion: We show that a MCT diet improves survival and temperature control during F+C due to improved shivering thermogenesis, but we also show that a MCT diet increases ventricular arrhythmias in mice with a VLCADD.

P-6

Deletion in the cardiac troponin I gene, resulting in premature stop codon, causes restrictive cardiomyopathy

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Introduction and objective: In contrast to dilated and hypertrophic cardiomyopathies, where over ten disease-causing genes have been identified and well described, mutations in only two genes, desmin and cardiac troponin I (TNNI3) have been shown to cause restrictive cardiomyopathy (RCM). Our aim was to apply a candidate gene approach and search for possible TNNI3 and desmin mutations in patient with RCM.
Methods and results: Direct DNA sequencing of the desmin and TNNI3 genes in a young patient with restrictive cardiomyopathy were performed. In cardiac troponin I gene we identified a novel one nucleotide deletion, resulting in frame shift and predicted formation of a premature stop codon, deletion of part of exon 7 and all exon 8, and truncation of significant C-terminal portion

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of TNNI3. Western blot analysis showed approximately 50% reduction of total troponin I content in myocardial tissue. The clinical hallmark was a restrictive type of cardiac hemodynamics, and progressive congestive heart failure, leading to the death of the patient at the age of 28.
Conclusion: We present a second report showing the involvement of TNNI3 in development of RCM, in this case as a result of frame shift with formation of a predicted stop codon and large deletion of the protein.

P-7

Computational/mathematical modeling of the systemic and pulmonary circulations with and without ventricular septal defect

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Objectives: To validate a computational/mathematical model of systemic and pulmonary circulations, with and without ventricular septal defect (VSD).
Methods:
–Normal heart. The electric/hydraulic analogy provided numerical simulation for pulmonary and systemic circulations, with heart chambers as active components, systemic circulation (aorta, arterial and venous tree, caval veins) and pulmonary circulation (pulmonary artery, capillaries and pulmonary veins) as passive components. Pressures, flows, hydraulic resistance and oxygen saturations were entered in the computational/mathematical model for the theoretical subject, a neonate 3.5 kg of body weight and 0.33 m^2 of body surface area (BSA).
– VSD. An unrestrictive VSD was added to the computational/mathematical model, and related changes of pressures, flows, hydraulic resistance and oxygen saturations were numerically computed.
– Validation. Numerical results for the "VSD" model were compared with data obtained from cardiac catheterization in 8 patients with VSD, mean age 5.9 ± 2.5 months, mean weight 5.4 ± 1.2 kg, mean BSA 0.34 ± 0.08 m^2.
Results:
–Normal heart. Pressure/volume loops obtained for right and left ventricles, pressures, flows and resistance curves and oxygen saturations for right and left ventricle, aorta and pulmonary artery, were similar to reported physiological data adequate for age and weight.
– VSD. No statistical differences were found between computational/mathematical model and clinical data for the ratio of right/left ventricular systolic pressures (0.10 vs 0.94 ± 0.06, N.S.), pulmonary artery/aorta pressures systolic (0.10 vs 0.92 ± 0.08, N.S.), diastolic (0.58 vs 0.60 ± 0.02, N.S.) and mean (0.85 vs 0.85 ± 0.01, N.S.), oxygen saturation in aorta (98 vs 98.4 ± 0.7%, N.S.), superior vena cava (65.5 vs 67.9 ± 1.6%, N.S.) and pulmonary artery (84.2 vs 84.5 ± 3.6, N.S.), QP/QS (2.4 vs 2.4 ± 0.8, N.S.) and PVR/SVR (0.4 vs 0.4 ± 0.1, N.S.).
Conclusions: The computational/mathematical model provides a validated calculation of the hemodynamic parameters and oxygen saturations in pulmonary and systemic circulations, with and without VSD. The application of the validated model can be extended to other congenital heart defects with associated malformations, as well to study the effects of interventional and/or surgical procedures on pulmonary and systemic circulations.

P-8

Evaluation of 4 Methods of DNA Extraction from Formalin-Fixed, Paraffin-Embedded Human Heart Samples

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Introduction: It is possible to extract DNA from archival tissues by different methods but so far these methods were only compared two-by-two and no study was undertaken to test the quality of extracted DNA.
Aim: Four methods of DNA extraction of archival tissue were compared and the sequence of extracted DNA was analysed.
Methods:
–DNA extraction: fresh human tissue specimen was cut in two samples: one was frozen and the other one was formalin fixed and paraffin embedded. DNA was extracted from formalin/paraffin slices using 4 different methods (1) tween 20/chelex 100, (2) triton X100, (3) Proteinase K, (4) heat. The frozen sample used as control was extracted by the salting out method.
DNA testing: PCR amplification of a 200-nucleotide (nt) fragment and nested PCR amplification of a 529-nt fragment were performed on DNA extracted with the 4 methods. Furthermore, a fragment of 366 nt was PCR amplified and cloned. Several clones were sequenced and insert sequence was compared to the control sequence.
Results: The success rate of the 200-nt fragment amplification was 75%, 0%, 58% and 90% for methods 1 to 4, respectively. The success rate of the 529-nt fragment amplification by nested PCR was 25%, 25%, 0% and 90% for methods 1 to 4, respectively.
— DNA extracted by methods 1 and 4 was further studied.
  — Seven different polymorphic markers were tested on DNA extracted from formalin/paraffin and frozen samples of the same human heart. No allelic variability was observed between frozen and fixed samples whether DNA was extracted by methods 1 or 4.
— Ten clones obtained after PCR amplification of a 366-nt fragment from DNA extracted from frozen and fixed heart tissue (methods 1 and 4) were sequenced. The mutation rate of DNA from frozen sample was 5×10^{-4} compared to 20×10^{-4} from fixed tissue (methods 1 and 4).
Conclusions: Extraction of DNA from formalin-fixed, paraffin-embedded heart tissues is feasible by tween 20/Chelex 100 and heat methods. The four-fold increase in mutation rate after fixation should not induce misleading sequence variations although these sequence variations tend to occur at particular positions.

P-9

Correlation between PTPN11 gene mutation and congenital heart defect and the ECG pattern in Noonan syndrome

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Introduction: Noonan Syndrome (NS) is a developmental disorder mainly characterized by facial features, short stature and a congenital heart defect. The most common described heart defect is pulmonary

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stenosis (PS) followed by hypertrophic cardiomyopathy (HCM) and Atrial septal defect (ASD). The electrocardiography (ECG) of NS patients often displays a characteristic pattern, which consists of a wide QRS complex and/or left axis deviation and/or abnormal Q wave and/or little r development over the left precordium. In 2001 a mutation in the PTPN11 gene on chromosome 12q24.1 was found to be a major cause for NS.

Methods: We evaluated the presence of a congenital heart disease and ECG in NS patients in whom PTPN11 gene mutation analysis was performed.

Results: In this study, 84 NS patients were included, 56 of them had a PTPN11 mutation. PS was the most common heart defect (61.9%), ASD was present in 10.7% and HCM in 8.2%. A statistically significant correlation was found for PS with PTPN11 mutation (71.4% versus 42.9%; $p = 0.017$). A significantly higher prevalence of HCM was observed in the group without a PTPN11 mutation (21.4% versus 1.8%; $p = 0.005$).

At least one characteristic ECG feature was present in 50% of the NS patients. Left axis deviation was present in 38 of 84 (45.2%) NS patients, little r development over the left precordium in 20 of 84 (23.8%) patients and a pathologic Q in 5 of 84 (6.0%) patients. A wide QRS complex was not found in any of these patients. No significant difference was found in ECG characteristics between PTPN11 mutation positive and negative NS patients and NS patients with and without a (particular) heart defect.

Conclusion: Pulmonary stenosis is the most common congenital heart defect in our patients with Noonan syndrome, and is significantly associated with the PTPN11 gene mutation.

However the characteristic ECG pattern is neither related to the PTPN11 gene mutation, nor to the presence of a (particular) heart defect

P-10

Intravenous tezosentan and vardenafil equipotently reverse pulmonary vasoconstriction in experimental normobaric hypoxic pulmonary hypertension

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Introduction: Excessive hypoxic pulmonary vasoconstriction constitutes the basis of maladaption to high altitude that can lead to pulmonary edema, right heart failure and death. Increased phosphodiesterase activity as well as increased levels of endothelin-1 have been addressed as causers of increased pulmonary vascular tone.

Objective: To define the hemodynamic and intrapulmonary effects of tezosentan, which is an intravenous dual endothelin-A (ET_A) and endothelin-B (ET_B) receptor blocker, and vardenafil, which is a novel type phosphodiesterase-5 (PDE-5) antagonist, that shows more potent and selective inhibition of PDE-5 than sildenafil, in a pig model of normobaric hypoxic pulmonary hypertension.

Methods: Twenty-four 4 weeks old ventilated white farm pigs were exposed to normobaric hypoxia (FiO_2 12%) and randomly assigned to four groups per six animals each in order to receive either intravenous tezosentan, vardenafil, combined tezosentan and vardenafil or to serve as control.

Results: Hypoxia led to a mean 70% increase of mean pulmonary artery pressure (Ppa) and to a mean 112% increase of pulmonary vascular resistance index (PVRI). After 90 minutes of treatment with either tezosentan or vardenafil Ppa and PVRI were significantly lower in the treatment groups than in controls ($p < 0.001$ for both parameters), without affecting arterial blood pressure. Cardiac index increased significantly after induction of hypoxia only in those animals, receiving vardenafil ($2.81 \cdot min^{-1} \cdot m^2 \pm 0.7$ to $4.21 \cdot min^{-1} \cdot m^2 \pm 0.7$, $p = 0.0003$). Ventilation-perfusion matching was not significantly altered by either tezosentan or vardenafil.

Conclusion: Intravenous tezosentan, as well as vardenafil equipotently reverse pulmonary vasoconstriction without afflicting pulmonary gas exchange. Vardenafil, but not tezosentan increases cardiac index in experimental hypoxic pulmonary hypertension.

P-11

Maternal Ingestion of Green Tea, Mate Tea and Grape Juice Cause Fetal Ductal Constriction: an Experimental Study

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Background: The role of maternal ingestion of nonsteroidal anti-inflammatory drugs in triggering fetal ductal constriction, a predisposing factor for neonatal pulmonary hypertension, is related to inhibition of prostaglandins. Herbal teas widely used in pregnancy, such as green tea and mate tea, as well as grape juice, have polyphenol components, especially 3-gallate-gallocatechin and resveratrol, with antioxidant and anti-inflammatory effects depending on inhibition of cyclooxygenase-2. This experimental study was designed to test the hypothesis that maternal consumption of green tea (GT), mate tea (MT) and grape juice (GJ) causes fetal ductal constriction.

Methods: Thirteen near term fetal lambs (>120 days) were assessed by fetal Doppler echocardiography (FDE) with color flow mapping before maternal administration of concentrated dosages of GT to four sheep, MT to four and GJ to five as the only source of liquid. After 1 week, a control FDE was performed in the survivors. Morpho-histological analysis was carried out in the fetal specimens who died after exposure. Differences were compared by Wilcoxon paired test.

Results: In seven survivor fetuses (3 exposed to GT and 4 to MT), inequivocal evidences of constriction of ductus arteriosus were demonstrated, with increase in mean ductal systolic velocities (0.80 ± 0.19 m/s to 1.17 ± 0.15 m/s, $p = 0.018$), mean diastolic velocities (0.21 ± 0.05 m/s to 0.31 ± 0.01 m/s, $p = 0.018$) and mean right to left ventricular dimension ratios (1.05 ± 0.14 to 1.43 ± 0.23, $p = 0.02$), as well as ductal flow turbulence, leftward ventricular septal bulging and tricuspid regurgitation in all. Pulmonary regurgitation was present in 2 fetuses. There was 1 fetal death in the group receiving GT. All 5 sheep exposed to GJ died of acidosis after 2 and 3 days. Autopsy in the 6 fetal specimens showed enlarged right ventricles and histological evidences of ductal constriction (decrease in ductal/pulmonary artery inner diameter ratio and gross increase in periluminar avascular zone thickness).

Conclusion: This experimental study provides evidences that maternal ingestion of the polyphenol-rich beverages green tea, mate tea and grape juice at late gestation trigger constriction of fetal ductus arteriosus. This knowledge should pompt preventive actions in perinatal dietary orientation.

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P-12

Left ventricular aneurysm in fetal life – a rare entity with an unpredictable outcome

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Introduction: Congenital left ventricular aneurysm (CLVA) is a very rare malformation, with still unclear etiology and a variable prognosis. Around 400 cases are reported in the literature, 31 with presentation in utero, more than 1/3 dying and early detection is considered a negative prognostic sign. We present two cases of CLVA, diagnosed at a similar gestational age, aiming to contribute to the general knowledge of this particular anomaly.
Case 1: A 33 years old healthy woman at 2nd pregnancy was referred at 21 weeks' gestation (w.) for fetal echocardiogaphy and apical aneurysm of the LV was found, of 15mm diameter, with thin hypokinetic walls, normal mitral and aortic flow and normal heart rate, in good compensation. Viral check-up was negative. At 25 w.g. the whole LV was markedly dilated (21×34mm), akinetic, with mild mitral and aortic regurgitation, functional aortic atresia and ascites. At 26 w.g. intrauterine death occurred and autopsy confirmed the echographic findings, with normal coronary arteries.
Case 2: A 27 years old healthy woman, primigravida was referred at 21.4 w. for fetal arrhythmia and "strange" LV. A submitral aneurysm extended till the apex was found (mid-LV diameter 11mm, normal LV length, kinesis, mitral and aortic flow) and persistent supraventricular bigeminy-trigeminy. Check-up for infections was negative. At follow-up the fetus remained compensated, with growing dimensions of the aneurysm, elongated apex, thin hypokinetic LV posterior wall (diam. 21×31 mm, SF 6–15%), normal mitral and aortic flow and variable extrasystolic arrhythmia. A male baby (2.5 kg) was born by Cesarean section at 38 w.g, in good condition, with extrasystolic bigeminy-trigeminy, SF 17% at the level of aneurysmatic portion, 29% at the level of mitral valve. Treatment with furosemide and captopril was started controlling well the heart compensation; after a few days, the aneurysmatic dilation further increased with bulging of the mid-portion of the septum, and a small secondary diverticulum of the apex. The baby is now 11 months old and well, with rare ectopic beats.
Conclusions: Our two cases show the difficulties in predicting the prognosis of CLVA. A straight follow-up in utero is mandatory to indicate an evolutive tendency.

P-13

Diastolic function of the fetal heart assessing by tissue Doppler imaging: a comparison to premature infants and neonates

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Introduction and objectives: The fetal heart has unique diastolic function because of its immaturity and specific fetal circulatory conditions. After the birth, significant changes in diastolic filling pattern toward adult profile became evident. The aim of this study was to analyze myocardial velocities of the fetal heart in the last trimester of the pregnancy and to compare this data with results in the premature infants (of similar gestational age) and neonates in the first few weeks of life.
Methods: Tissue Doppler echocardiography was performed in 35 healthy fetuses ranging 27–37 weeks (mean 30.17 ± 2.79 weeks) and compared with data measured in 32 premature infants (born after mean gestational age of 33.09 ± 1.71 weeks) at the age of 1–21 days (mean 8.56 ± 5.15 days) and 31 neonates of the mean age of 12.9 ± 5.83 days (ranging 3–24 days).
Results: The main differences were in Em velocity and Em/Am ratio at the both ventricles. Left ventricle Em was significantly higher in the neonates (mean 10.55 cm/s) then in other two groups (mean 7.14 cm/s in fetuses, 8.09 cm/s in preterm babies, $p < 0.0001$), and a similar findings were for Em of the RV. Em/Am ratio was significantly higher after birth but the mean values didn't exceed 1 (mean values for the LV were 0.62 in fetuses, 0.82 in premature infants and 0.91 in neonates, $P<0.0001$; mean values for the RV were 0.62, 0.63 and 0.8, respectively, $P<0.004$). Mean Em/Am ratios were significantly lower than mean E/A ratio calculated from the PW Doppler in the preterm infants (0.82 vs. 1.11 for the LV, $P < 0.0001$, and 0.67 vs. 0.87 for the RV, $P<0.001$) as well as for the LV in neonates (0.91 vs. 1.06, $P<0.009$). Tei index was almost the same in all groups for both ventricles, ranging 0.48–0.53.
Conclusions: Ventricular filling pattern are specific and different in fetal period, and alteration occurred after the birth are consequences of remarkable changes in loading conditions, as well as myocardial maturational changes.

P-14

Decreased serum insulin-like growth factor-I and insulin-like growth factor binding protein-3 associated with growth failure in patients with cyanotic heart disease

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Objectives and methods: Cyanotic congenital heart disease in children commonly causes more pronounced growth retardation in comparison with acyanotic congenital heart disease. To determine whether chronic hypoxemia results in alterations in endocrine function that may contribute to growth failure, we measured serum insulin-like growth factor I (IGF-I), insulin-like growth factor binding protein-3 (IGFBP-3) in 40 cyanotic (22 male, 18 female, age = 6.3 ± 5.1 years) and 22 acyanotic (11 male, 11 female, age = 8.2 ± 3.7) patients with congenital heart disease. Growth and nutritional status of each patient was determined by using anthropometric parameters and calorie and protein intake assessments.
Results: Although the calorie intake in cyanotic patients was not significantly different from the acyanotic ones and protein intake was higher than those, standard deviation of mid arm circumference (MAC) (−1.99 ± 1.1 vs −0.48 ± 1.0, $p = .000$), triceps skinfold thickness (TST) (-1.76 ± 0.9 vs -1.0 ± 1.1, $p = .003$), body mass index (BMI) (-1.72 ± 1.6 vs -0.77 ± 1.1, $p = .006$) as well as weight for height (90 ± 13 vs 98 ± 14, $p = .026$) values were significantly lower in cyanotic patients than in acyanotic ones. In addition, standard deviation of serum IGF-I (−2.71 ± 0.8 vs −1.1 ± 1.9, $p = .000$) and IGFBP-3 (−1.93 ± 1.2 vs 0.05 ± 1.5, $p = .000$) concentrations were also significantly reduced in cyanotic patients.
Conclusions: In the light of these results it can be thought that the main factor that leads to growth failure in cyanotic patients may be the deterioration in endocrine function due to a decrease in somatomedins, rather than undernutrition.

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P-15

Clinical spectrum, morbidity, and mortality in 41 pediatric patients with ventricular noncompaction

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Introduction: Ventricular noncompactin is a congenital pathological entity that can occur in isolated form or associated with other heart disease and often involves both ventricles. The purpose of this study was to identify the clinical features, morbidity, mortality, and prognosis of children with isolated NC and congenital heart disease associated with NC.
Methods: We retrospectively reviewed 41 patients with ventricular NC evaluated.
Results: There were 23 girls and 18 boys. The median age at presentation was 3.2 years (range: 1 day 17 year). Twenty-two of the 39 (54%) patients were diagnosed under the 2 years of age. On the physical examination, finding of cardiac failure, murmur, failure to thrive, cyanosis and dyspnea was detected.
Electrocardiographic abnormalities were marked in 30 patients. In echocardiographic assessment, NC morphology was found in the left ventricle (LV) in 19 patients, in right ventricle (RV) in 10 patients and in both ventricles in 10 patients.
Among the total 14 patients with complex cardiac pathology; nine had biventricular NC, 3 had RVNC and the remaining 2 had LVNC.
Among 8 patients with ASD or/and VSD, 6 were associated with LVNC, whereas 2 with RVNC. Three had pulmonary stenosis all had RVNC. Among 12 patients with cardiomyopathy diagnosis, 5 had dilated cardiomyopathy (4 LVCN and 1 had just RVNC). Rest 7 of these 12 diagnosed with LVNC. Eleven patients with complex cardiac pathology were operated. During postoperative period 8 of them died. Two remaining patients were also lost due to infections and one patient was applied stent due to pulmonary atresia. Among the 16 patients diagnosed as isolated NC, 4 still wait in the cardiac transplantation list.
Conclusion: Congenital heart disease associated with both ventricular noncompaction are indicators of poor prognosis. However heart failure, arrythmia, embolism and sudden death are common clinical findings in isolated NC. Althgough etiology of NC is not fully eluciated its diagnosis is possible since birth or in fetal life. Echocardiography is the golden standard in diagnosis and early diagnosis is crucial in clinical progress of these patients.

P-16

Vascular endothelial growth factor, IL-6 and IL-8 levels in congenital heart disease

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Objective: Increased pulmonary flow in congenital heart diseases with left to right shunt lesions and hypoxia in the congenital heart diseases with right to left shunt lesions constitute the main course cause of the morbidity and mortality. We aimed to determine the role of vasoactive mediators in morbidity of congenital heart diseases.
Methods: Patients (31 F, 22 M; 5 months–16 y, mean: 4.31 ± 4.19y) were divided into two groups according to their shunt lesions: acyanotic group 1 (group 1a: pulmonary artery systolic pressure >30 mmHg and group 1b <30 mmHg) and cyanotic group (Group 2). All patients were divided into two groups according to their pulmonary artery systolic pressure (group PA 1: pulmonary artery systolic pressure <30 mmHg and group PA 2 >30 mmHg) and pulmonary resistances (Group Rp1: pulmonary resistance <2 Ü/m^2 and Group Rp2: >2 Ü/m^2).
VEGF, IL-6 and IL-8 levels were determined in all patients.
Results: The VEGF level (pg/ml) was increased insignificantly in the acyanotic group (219.2 ± 32.8)vs cyanotic group (125.2 ± 207.1), and Group 1a (300 ± 425) vs Group 1b (117 ± 93). In Group PA2VEGF levels (244.8 ± 374) were higher than Group PA1 (132.0 ± 173) $p > 0.05$.
VEGF levels were significantly higher in Group Rp2 (320.8 ± 411.6) than Group Rp1 (102.8 ± 128.9) $p < 0.05$.
The IL-6 level (pg/ml) was increased in Group 2 (38.3 ± 128.0 vs 4.09 ± 6.12), Group 1a (5.43 ± 8.02 vs 2.41 ± 0.99), Group PA2 (27.6 ± 103.4 vs 2.62 ± 1.70), $p > 0.05$.
The IL-8 level (pg/ml) was increased in Group 2 (44.1 ± 68.1 vs 27.0 ± 68.4), Group 1a (43.4 ± 89.5 vs 6.42 ± 4.91), Group PA2 (48.8 ± 83.3 vs 14.10 ± 35.07) $p > 0.05$, and Group Rp 2 (43.8 ± 89.31 vs 7.72 ± 7.51) $p < 0.05$.
Positive correlations were found between VEGF and IL6, IL8, pulmonary artery systolic pressures. Arterial O_2 saturations showed negative correlations with VEGF and IL8 levels in the Group 1.
Conclusion: According to our results pulmonary artery resistance has major effect on VEGF levels rather than hypoxia. VEGF plays an important role in the development of irreversible pulmonary hypertension; therefore evaluation of VEGF levels could be guiding in the management of treatment.

P-17

A Large Pedigree With Mild And Severe Cases Of Congenital Heart Defects Segregates A Zic3 Missense Mutation

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Introduction: It is assumed that about 80% of congenital heart defects (CHD) are secondary to an oligenic type of inheritance with a major genetic factor that can be modulated by other minor genetic and environmental factors. Families with multiple cases of CHD provide an opportunity to decipher these genetic factors.
Method: A National Registry was started in 2003 to collect families with multiple cases of CHD. Twenty seven paediatric cardiologists are participating. Up to now, 97 families were enrolled including 234 affected and 399 non-affected individuals. A blood sample was obtained from 60% of these individuals. A systematic screening for mutation was carried out in 3 genes (GATA4, NKX2.5 and ZIC3) in the DNA of 2 affected individuals per family. This study was approved by ethical committee.
Results: A missense mutation was found in ZIC3 (p.Gln292His) in a boy with absence of inferior vena cava, polysplenia, intestinal malrotation, extrahepatic biliary atresia and mild bilateral talipes equinovarus. He had surgery (Kasaï intervention). This mutation was found in his heterozygous mother. She has no heart malformation (ECG and echocardiography) but mild speech difficulties. She is part of a large sibship with 9 girls and only 1 male. One of the maternal aunts of the proband has an ASD, a first degree AV block and a left anterior hemiblock. This woman has a single boy with

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ostium secundum ASD. The single maternal uncle has also an ASD. Finally, a remote cousin had anomalous venous return, mitral and aortic atresia. This complex CHD was disclosed at birth and he died shortly after birth. A blood sample was taken from him and his parents. Collection of blood samples from the large sibship is underway so that the mutation carrier status is tested soon.
Conclusions: A mutation in ZIC3 is presumably responsible for cardiac and extra-cardiac defects that are severe in males and mild in females. The ZIC3 mutation could also account for the male/female imbalance in the large sibship (1/8) if hemizygous mutation in males is frequently lethal. A complete account for ZIC3 mutation in this family will be reported at the AEPC meeting.

P-18
Hospitalization for Respiratory Tract Infection in Young Children with Hemodynamically Significant Congenital Heart Disease: The 'CIVIC II' Study, Spain 2005–2006
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Objectives: Second seasonal evaluation on hospitalization rate for acute respiratory tract infection in children less than 24 months with hemodynamic significant congenital heart disease, secondary objectives associated risk factors, preventive measures, aetiology and clinical course.
Methods: 764 subjects were followed from October 2005 to April 2006 in an epidemiological, multicenter (53 Spanish hospitals), observational (descriptive), follow-up, prospective study.
Results: 100 patients (13.1%, 95% CI: 10.9–15.7%) required a total of 120 hospital respiratory infection-related admissions. Significant associated risk factors for hospitalization included:

Risk Factor for Hospitalization for RI	Risk	Lower limit (CI95%)	Upper Limit (CI95%)
Age younger than 12 months	4.2	2.0	8.3
Siblings less than 11 years old	2.8	1.7	4.4
Previous respiratory disease	2.4	1.2	4.9
Sex (Male)	2.1	1,3	3.3
Previous Surgery with cardiopulmonary bypass	2.0	1.1	3.4
Weight under 3rd percentile	1.9	1.2	3.1

Bronchiolitis (41.7%), upper respiratory tract infection (31.7%), and pneumonia (17.5%) were the main diagnoses. An infectious agent was found in 33 cases (27.5%): 21 respiratory syncytial virus, 3 haemophilus influenzae, 3 stafiloccocus, 2 streptoccocus pneumoniae, 2 adenovirus, 2 varicela, 2 pseudomona. The odds ratio for respiratory syncytial virus infection hospitalization increases by 1.8 in patients with incomplete respiratory syncytial virus prophylaxis. The median length of hospitalization was 8.5 days. In 25 episodes admission in intensive care unit and in 9 cases mechanical ventilation was required. 4 patients died from respiratory infection.
Conclusions: In a second season, hospital admissions for respiratory infection in young children with hemodynamic significant congenital heart disease are mainly associated with non-cardiac conditions (age, siblings, respiratory disease, sex, malnutrition) and cardiopulmonary bypass. Bronchiolitis due to respiratory syncytial virus was the most commonly clinical picture and identified infectious agent. Incomplete propyilaxis increased hospitalizations.

P-19
Can Preoperative Testing Predict Postoperative Outcome?
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Introduction: Evaluating pulmonary vascular resistance by testing pulmonary vasoreactivity has become a useful tool in determining the suitability for corrective surgery or interventional procedures. The purpose of this study was to define the predictive power of preoperative testing in regard to outcome after corrective surgery/intervention.
Methods: All testing protocols performed at our institution from 1998 to 2006 were reviewed; those of patients with simple intracardiac shunting defects under consideration for corrective surgical/interventional procedures were further analysed. Pulmonary vasodilation was achieved with oxygen, nitric oxide, prostanoids, or combinations thereof. Systemic and pulmonary artery pressures were assessed and systemic and pulmonary vascular resistances and blood flows determined using the Fick principle. Outcome was defined as poor (death, right heart failure or pulmonary hypertensive crisis in the postoperative interval) versus good.
The predicting value of testing results regarding outcome was evaluated using calculations of sensitivity, specificity and comparison of receiver operating characteristic (ROC)-analysis under different test conditions.
Results: Out of 214 pharmacological testing procedures, 82 were performed to determine operability, which led to surgical (n = 52) or interventional (n = 9) procedures in 61 cases. No differences in pulmonary vasodilation in the used substances were seen. 13 patients had a poor outcome with two late deaths after 5 and 6 months. Rp:Rs at baseline (Rp1) and Rp:Rs at maximum vasodilation (Rp2) were higher in patients considered operable vs inoperable, and also in those with poor vs good outcome. Comparison of ROC-Curves for Rp1 and Rp2 did not show significant differences, providing an optimal balance of sensitivity and specificity for Rp1 < 0.60 and Rp2 < 0.30 as potential criteria for operability.

	inoperable	operable	poor outcome	good outcome	ROC
Rp1 (base)	1.14 ± 0.24	0.37 ± 0.03	0.57 ± 0.08	0.32 ± 0.03	<0.60
Rp2 (test)	0.66 ± 0.18	0.18 ± 0.02	0.27 ± 0.04	0.16 ± 0.02	<0.30
	p < 0.05	p < 0.05	p < 0.05	p < 0.05	p < 0.05

Conclusions: In our patient cohort, predictive power was not very strong, but consistent, indicating that an Rp:Rs <0.30 measured at maximum pulmonary vasodilation as a criterion for operability would have been the best value to predict a good outcome while preventing an untoward one. Our results suggest, however, that also an Rp:Rs < 0.6 measured at baseline had the same predictive power.

P-20
Waist circumference and early detection of Metabolic syndrome
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According to recent investigations, metabolic syndrome is considered as a predictor of cardiovascular diseases (CVD) and type 2 diabetes. Early recognition of the syndrome or its components may help to decrease CVD morbidity and mortality rates in population.
Purpose of the study was to investigate association of waist circumference (WC) and some components of metabolic syndrome (blood lipid levels, arterial blood pressure(BP)) and anthopometrical parameters in adolescent population of Novosibirsk.
Methods: In 2003 the representative sample of the Novosibirsk adolescents of both sexes aged 14–17 (667 subjects: 301 boys, 366 girls) were examined. The program included questionnaire, double measurement of arterial blood pressure, anthropometry, (weight, height, body mass index, circumferences for chest, waist, hips, subscapular skinfolds and triceps skinfolds), assessment of blood lipid levels (total cholesterol, triglycerides, high-density lipoprotein cholesterol (HDL-C)). Adolescents were divided into 3 groups: non-obese (BMI < 85 percentile) - 567 (84.7%), overweight (BMI 85–94 percentile) – 69 (10.3%), and obese (BMI ≥95 percentile) –33 (4.9%). In each group the average values of the parameters were evaluated.
Results: Mean systolic and diastolic BP, triglycerides and anthopometrical parameters of the obese adolescents were higher and levels HDL-C were lower comparing to the non-obese and overweight adolescents (P≤0.001).
Associations (P≤0,001) between WC systolic BP (boys r = 0.42, girls r = 0.28), triglyceride level (boys r = 0.20, girls r = 0.17), HDL-C (boys r = −0.19, girls r = −0.14), BMI (boys r = 0.89, girls r = 0.80), height (boys r = 0.40, girls r = 0.21), weight (boys r = 0.87, girls r = 0.79), chest circumferences (boys r = 0.87, girls r = 0.81) and hips' circumferences (boys r = 0.76, girls r = 0.67) were found in both gender groups.
Multiple linear regression analysis showed the WC as a better independent predictor for the systolic BP (P≤0.001), HDL-C (P≤0.001), triglycerides (P = 0.012), than the BMI (P = 0.029, P = 0.33, P = 0.25 accordingly).
Conclusion: Waist circumference may be useful as an early predictor of metabolic syndrome in adolescents.

P-21
Utility of a regional campaign for early diagnosis and treatment of Kawasaki's disease to prevent cardiac-coronary complications
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Background: Kawasaki disease (KD) is an acute self-limited vasculitis of childhood characterized by fever, bilateral conjunctivitis, erythema of the lips and oral mucosa, changes in the extremities, rash, and cervical lymphadenopathy. Coronary artery aneurysms or ectasia develop in 15% to 25% of untreated children and may lead to ischemic heart disease or sudden death.
Methods and Results: This study was carried out in the Marche Region, Italy (population 1.500.000 inhabitants). For 2 years, since 1984 to 1986 was performed by Lancisi's Hospital cardiologic staff a regional campaign to make aware Paediatricians of the complications of KD, having had in different Hospitals of the Region 3 cases of death for infarction in the first years of 80's. No devoted budget was used. Local meetings with general and hospital pediatricians were planned. An early access to the echo lab of the our Center was offered in emergency to all the paediatric regional community for the clinical suspicion of KD. Until now the cases of confirmed diagnosis of KD are 218. Coronary compications: 35%. Major complications: 0.4%. Moderate complications: 0.8%. Mild involvement >30%. No cardiac complications: 65%. Complications were more than in literature, probably due to the more frequent use of echo in earlier stages. Diagnosis of KD was so made in cases previously judged negative. More pts were so treated according to the international guidelines. Long-term management of patients with KD was tailored to the degree of coronary involvement.
Conclusions: After the campaign, from 1986 until now in the Marche region was not signalled a single death or myocardial infarction episode due to KD. Our data show that KD complications were detected more frequently than in other studies but the outcomes of this population were really good and significantly better than in most literature data. This outcome improvement was reached whithout any financial resources after only two years of local unexpensive meetings but by the offering of early specialized point of diagnosis to paediatricians.

P-22
The Kurdistan Experience: Creating Paediatric Cardiac Services in the post-Saddam Era
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Introduction: In the Iraqi Kurdistan Region (IKR), emerging with difficulty from years of persecution, healthcare is in a state of extreme deterioration. Management of children with congenital heart disease (CHD), roughly estimated as 4000 out of a population of approximately 4 million, has become an emergency. In 2006, the Mediterranean Institute of Hematology and Medchild (Italian non-profit foundations) with Policlinico San Donato (PSD) have started a pilot project aimed at: 1) assessment of children with CHD in IKR; 2) treatment of urgent cases in specialised Italian centres; 3) training healthcare professionals from IKR in diagnosis and management of CHD; 4) Optimising healthcare facilities in IKR for management of CHD.
Preliminary Report: 1) In 4 clinical missions, a team of professionals travelled to IKR and assessed in total 685 patients aged 0.1 to 38.5 yrs (mean 6.1), with known/suspected CHD. 626 were found affected by CHD needing treatment. 156 were deemed urgent, i.e. needing treatment within 6 months. Sixty-seven were considered inoperable. Three groups of 25, 29 and 15 patients, selected as urgent and with positive expected outcome, were transferred to Italy (PSD, Ospedale Bambino Gesù, Policlinico S. Orsola) for treatment. We estimated a mortality rate in waiting list of 1–2/month. 2) Sixty-five patients had cardiac interventions in Italy. Fifty-three had complete repair, 12 had a palliative procedure. Five patients had transcatheter procedures. Sixty-two patients were discharged in good condition. Fifty-seven have returned to IKR. One patient with complex CHD died shortly after surgery. Significant complications occurred in 2 cases. Two patients needed re-operation for residual disease. 3) Two cardiologists and 2 surgeons from IKR have been included in 6-month to 2-year training programmes in Italy. Training programmes for nurses are under development. 4) A centre with facilities for management of CHD is being developed in the city of Erbil, under the team supervision. Six echocardiographic machines were donated to clinic centres for CHD in the IKR territory.

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Conclusions: Preliminary results are encouraging and show that political stability and institutional co-operation are essential to succeed. This pilot project creates a framework that could be applied to other countries and regions, with participation of other European centres.

P-23

Comparison of clinical profile in children with different types of left ventricular hypertrophy in hypertrophic cardiomyopathy

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Background: There are two major types of LV hypertrophy in hypertrophic cardiomyopathy (HCM). However, there is no published study comparing clinical features of concentric HCM (cHCM) and asymmetric septal hypertrophy (ASH) in children. The aim of our study was to compare clinical variables in patients with cHCM and ASH.
Methods: Retrospective analysis of 53 pts (33 boys, 20 girls, mean age 10.9 ± 4.8 yrs) with HCM diagnosed from 1991 to 2006. ASH was defined as isolated interventricular septum hypertrophy (IVS thickness > mean normal range relative to body surface area). Patients with both: IVS and LVPW hypertrophy, were diagnosed as having cHCM. Patients demographics and clinical variables as well as the results of echocardiography, 12-leads ECG, chest X-ray, 24-hour Holter ECG, single photon emission computer tomography (SPECT), ambulatory blood pressure monitoring, exercise test were analyzed.
Results: Children with ASH and cHCM do not differ regarding: incidence of syncopes, pre-syncopes, chest pain, palpitations, cardiac arrest, heart failure, family history of HCM and SCD. Children with ASH were older at diagnosis than pts with cHCM (80.0 ± 74.8 vs 41.3 ± 47.4 months, p = 0.027), less often had symptoms such as fatigability, effort dyspnea (6.25% vs 28.6%, p = 0.047) and RV hypertrophy observed in ECG (6.25% vs 28.6%, p = 0.047). Frequency of LV hypertrophy in ECG was similar in both groups (59.4% vs 85.7 p = 0.07); however Sokolov index was significantly higher in concentric HCM (34.9 ± 155 vs 61.4 ± 39.6 mm, p = 0.008). Echocardiography results are presented in the table:

Parameter	ASH	Concentric HCM	p value
IVS (% of mean normal range)	222.0 ± 72.2	316.5 ± 133.2	0.006
LVPW (% of mean normal range)	104.7 ± 16.5	199.0 ± 57.2	<0.001
LVOTO (% of pts)	34.4%	66.7%	0.027
LVOTO (gradient mmHg)	8.1 ± 16.8	42.7 ± 40.0	0.001
Aortic insufficiency (% of pts)	3.3%	52.9%	<0.001

Conclusions: (1) Patients with two different types of HCM share many similarities, although children with concentric HCM were diagnosed at younger age and more often presented with fatigability. (2) In pts with concentric HCM aortic insufficiency and LVOTO were more frequent and obstruction to flow was higher.

P-24

Metabolic cardiomyopathies in a paediatric population: clinical spectrum and outcome

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Introduction: Cardiomyopathies (CMP) may be idiopathic or secondary to an underlying definable systemic disorder, especially in a young population. A number of inherited metabolic disorders (IMD) will lead to hypertrophy or dilated cardiomyopathy. Identifying the metabolic aetiologies of CMP is important for genetic counselling and identification of appropriate therapeutic interventions.
Objective: To assess the clinical evolution of patients with cardiomyopathy secondary to IMD followed in a paediatric hospital.
Material and methods: The authors analysed the clinical records of patients with cardiomyopathy secondary to IMD and studied the following variables: sex; age of presentation, metabolic investigation; type of cardiomyopathy, treatment, time of follow up and outcome.
Results: The study included 12 children, 5 girls, aged 8 days to 13 years at the time of diagnosis of the metabolic disease. Metabolic, cardiac diagnosis and mortality are represented in table I:

Table I. Metabolic and cardiac diagnosis and outcome

	HCMP	DCMP	Death
Mitochondrial disorders (n = 6)	3 1⋆	2 1⋆	2 (DCMP)
LCHAD (n = 3)	1	2	3 (1 HCMP; 2 DMCP)
Lisosomal storage disease (n = 2)	2		
Glycogen storage disease (n = 1)	1		

⋆In one case we verified the evolution of HCMP to DCMP;
HCMP-hypertrophic cardiomyopathy; DCMP-dilated cardiomyopathy

Cardiomyopathy was diagnosed before the first year of age in 5 children, in the neonatal period in one case, and preceded metabolic diagnosis in 5 cases (1 LCHAD; 4 mitochondrial disorders). One patient initially presented with hypertrophic cardiomyopathy that evolved to dilated. Five children died (9 days to 11 years), two during the first year (both had LCHAD), due to cardiac failure (n = 4) and sudden death (n = 1).
Comments: In this study we found a higher prevalence of hypertrophic cardiomyopathy that was present in all storage disease. Dilated cardiomyopathy was found in mitochondrial disorders and LCHAD and was associated with high mortality. Mortality was higher when the diagnosis was made in the first year of life (3/5). The diagnosis of cardiomyopathy preceded the diagnosis of metabolic disorder in 42% of cases. Most of these diseases have no curative treatment. However, in some cases an early diagnosis and specific treatment can have a favourable impact on the outcome.

P-25

Sexual life in male adolescents and young adults with congenital heart disease

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Introduction: The advances in the diagnosis and management of congenital heart diseases had as a result a considerable number of patients reaching the adolescent and young adult life.
Objective: The aim of this study was to investigate the influence of the heart problem on the sexual life of male patients.
Material and Methods: Data was obtained by the records of 158 male patients followed up during 10 years (1995–2005) time.
Results: The selected age group was between 15-35 years, mean age 25. Simple heart lesions had 98 (62%) patients, 21 (13.3%) had combinations, 35 (22.2%) complex heart diseases and 4

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(2.5%) very complex heart diseases. For the management, 65 (41%) patients were just followed up, 11 (8%) were managed with interventional methods and 82 (51%) underwent heart operations. All of those with simple heart lesions or combinations of simple heart lesions as well as the majority of those with complex heart problems had normal sexual life for their age, 130 (82.2). Fifteen (9.5%) patients had sexual relations without intercourse, whereas 13 (8.3%) patients with very complex heart problems like univentricular heart, pulmonary atresia, transposition of great arteries with postoperative problems like arrhythmias or chest deformities had no sexual life at all. The most of them complained of easy fatigability, appearance and psychological problems. There were also 6 (3.7%) patients with mental retardation. Two patients were engaged 3 married and 1 had a baby.
Conclusion : The majority of male patients with congenital heart defects have normal sexual life, whereas those with severe complex heart lesions have not, specially if the result of the operation is not successful.

P-26

Pulse-oximetry derived non-invasive Peripheral Perfusion Index as a possible tool for screening for congenital critical left heart obstruction: Comparison with reference values on 10 000 normal newborns

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Introduction: Peripheral perfusion index (PPI) derived from new generation pulse oximeter signals, has been suggested as a possible method to detect illness causing circulatory embarrassment. PPI is a relative number and varies between monitoring sites and from patient to patient as physiologic conditions vary. The ratio of the pulsatile component (arterial blood) to the non-pulsatile component (venous blood, bone, tissue, skin) corresponds to the amount of blood at that monitoring site and is termed PPI, and is calculated independently of the patient's oxygen saturation. PPI is a relative assessment of the pulse strength at a monitoring site and ranges between 0.02 to 20.00%. We aimed to establish the normal range of this index in healthy newborns, and compare it with newborns with duct-dependent systemic circulation.
Methods: A case-control study with 10 000 normal newborns and 9 infants with duct-dependent systemic circulation (LHOD-group) from our ongoing prospective pulse oximetry screening study in the region of Västra Götaland, Sweden. Single right hand (preductal) and foot (postductal) measurements of PPI with a new generation pulse oximeter (Masimo Radical SET, version 4) were obtained prior to the neonatal physical examination in all newborn nurseries in our region.
Results: PPI-values from newborns between 1 and 120 hours of age show an asymmetrical, non-normal distribution with median PPI-value of 1.70 and interquartile range 1.18 to 2.50. The 5th percentile = 0.70 and 95th percentile = 4.50. All infants in the LHOD-group had either pre- or postductal PPI below the interquartile range and 5 of 9 (56%) were below the 5th centile cut-off of 0.70 ($p < 0.0001$, Fisher's exact test). A PPI-value <0.70 gave an Odds Ratio for LHOD of 23.75 (95% CI 6.36–88.74).
Conclusion: Lower PPI-values than 0.70 may indicate illness and a value <0.50 (1st percentile) indicates definitely subnormal perfusion. PPI-values might be a useful additional tool for early detection of newborns with duct-dependent systemic circulation.

P-27

Frequency of diagnostic ECG-abnormalities in children of parents with familial hypertrophic cardiomyopathy

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Introduction: Studies from centres of adult cardiology have reported that between 75–95% of adults having echocardiographic features of hypertrophic cardiomyopathy (HCM), or carrying a disease-causing mutation, have pathological ECGs. Several different criteria have been proposed for ECG-screening for HCM, but they have all been evaluated in predominantly adult populations. The aim of this study was to establish how common ECG-abnormalities are in children and adolescents with familial HCM, and which ECG-criteria are most useful in childhood.
Methods: 65 consecutive children with a mean age of 10.6yrs (range 0–20yrs), that had one parent with familial HCM, and thus 50% risk of inheriting HCM, were screened by means of digital ECG and detailed echocardiography including tissue Doppler analysis. Echocardiography established 27 (42%) with pathological hypertrophy (echo-positive group), and 33 (51%) with normal echocardiography (echo-negative group), and these were compared by various proposed ECG-screening criteria for adult HCM, and ECG high-risk criteria for sudden death, namely limb-lead QRS-sum > 10mV. A further 5 children had wall thickness within normal limits but abnormal diastolic function or systolic hypercontractility and were suspected mutation carriers.
Results: The echo-positive group had significantly higher limb lead QRS-sum ($p = 0.006$), and 12-lead QRS-sum ($p = 0.017$) when compared with the echo-negative group, but no difference in Sokolow-Lyon index, QTc or QT-dispersion. Assessing individual criteria a 12-lead QRS-sum > 24mV was present in 48.1% of echo-positive individuals and 6.1% of echo-negative individuals ($p = 0.0002$) giving an odds ratio of 13.4 (95%CI 2.9–72.5), Q-waves >25% of R in two leads in 22.2% versus 0% ($p = 0.0059$), giving an odds ratio of 20.3 (1.1-379), Q-waves > 0.3mV in two leads was present in 37.0% as compared to 6.1% ($p = 0.0038$), odds ratio of 9.1 (1.8–46.5). The other criteria showed no significant differentiation, but 18.5% in echo-positive group fulfilled high-risk ECG-criteria in limb-lead QRS-sum. The best screening is achieved by combining 12-lead QRS-sum > 24mv and/or Q-waves >25% of R in two leads which identifies the echo-positive group with a sensitivity of 55.6%, a specificity of 93.9%, a positive predictive value of 88.2% and a negative predictive value of 72.1%.
Conclusions: A pathological ECG is less commonly present in children with familial HCM than in adults.

P-28

The QT and QTc intervals dispersion in sporting teenagers

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Objective: To evaluate the QT interval dispersion in adolescent athletes regarding on the type of physical effort.
Methods. Subjects: 51 sporting teenagers (14–17 years old) organized in 2 groups. First group: 25 endurance-trained athletes (runners, football-players) and the 2nd group: 26 strength-trained athletes (wrestlers, boxers). Control group: 20 teenagers in the same age group, without any sign of cardiac suffering. ECGs were assessed

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on all the patients, athletes and non-athletes and used to calculate QT interval in three successive cardiac ECG cycles, the QT interval dispersion (QTD) (the difference between maximum and minimum value of QT interval) and the QTc interval dispersion (QTcD) (Bazzet's formula).
Results: The average values of QTD and QTcD in the 1st and 2nd group were superior than the values in the control group but the difference is not statistic significant.

	1st group	2nd group	control group
QTD	43.54∓21.03 ms	48.23∓12.56 ms	35.88∓10.22 ms
QTcD	50.81∓19.34 ms	53.59∓17.21 ms	39.23∓14.81 ms

The highest values of QT interval were found in strength-trained sporting teenagers. The highest values of QTD and QTcD were found in sporting teenagers from the 2nd group that it might be possible to have a higher ventricular arrhythmia risk. There wasn't any case with QT interval value longer than the normal.
Conclusions: At side of other parameters ECG, it is useful research screening of the QT interval and QTc interval dispersion during periodic controls, like indicator of the risk of the ventricular arrhythmias at sporting teenagers.

P-29

Muscular Dystrophy and Cardiac Disease in Children

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Introduction: Muscular Dystrophies (MD) are a heterogeneous group of rare diseases. Cardiac involvement (CD) is a conditioning factor for morbidity and mortality. Early therapeutic intervention can change prognosis.
Purpose: Evaluate type, severity and outcome of CD in several forms of MD, in order to improve prognosis through rational cardiac follow-up.
Methods: Retrospective study including all MD patients with a follow-up longer than six months, admitted from January 1990 to June 2006. MD type, cardiac clinical profile, left ventricular dimension and function assed by echocardiogram and EKG abnormalities were evaluated and results between first and last visit were compared.
Results: The study group consisted of 23 patients (pts), aged 0.8–15 years (median 8 years). Duchenne MD was present in 10; six had congenital MD, three limb-girdle (LGMD), Becker MD two, one oculo-pharyngeal and another one a non-classified MD. At presentation 21 pts had a normal cardiac evaluation. Left ventricle was enlarged with poor contractile function in five (23%), four (80%) with MD Duchenne and one with LGMD, two of them with severe heart failure. EKG was abnormal in three of the five pts: left ventricular enlargement, left bundle branch block and repolarization abnormalities. Single supraventricular premature beats were found in one patient with non-classified MD.
At latest visit, (6 months–11 years; median = 4 years), 21 were free from CD. In the group with abnormal echocardiographic findings, three pts did not show progression of heart disease, one died in refractory heart failure and one was lost for follow-up. Meanwhile, two pts were added to the left ventricular dysfunction group: one Duchenne MD and one Becker MD. The patient with rhythm disorder went on sinus node dysfunction associated with conduction disorder, requiring pacemaker implantation.
Discussion and Conclusion: Results reflect the unique features of a rare disease group with mild clinical CD expression in paediatric patients. MD and CD prevalence are similar to other published data. However, the finding of new CD cases and one fatal case at midterm, both favours the need for early therapeutic intervention.

P-30

Assessment of Relationship Between Selected Tissue Doppler Imaging Parameters of Left Ventricular Function and Systolic Blood Pressure Load in ABPM in Children After the Successful Repair of Aortic Coarctation

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Arterial hypertension which is often observed in children after the surgical repair of aortic coarctation (CoA) may lead to left ventricular dysfunction because of increased afterload.
The aim of the study was an assessment of relationship between selected tissue Doppler imaging parameters of left ventricular function and systolic blood pressure load in Ambulatory Blood Pressure Monitoring in children after the successful repair of CoA.
Study group consisted of 32 children with the mean age 12.01 ± 4.24 years after the successful surgical repair of CoA. The mean age at the surgery was 3.84 ± 4.04 years and the mean follow-up 8.17 ± 3.29 years. Control group consisted of 34 healthy children.
In all children from study and controls groups the echocardiographic examination with TDI mitral annulus motion analysis, off-line assessment of strain and strain rate parameters and ambulatory blood pressure monitoring with 24-hours systolic blood pressure load (SPL) evaluation were performed. The following TDI parameters were assessed: systolic, early diastolic and late diastolic velocity of mitral annulus motion (S´, E´, A´) and ratio E´/A´. Of-line TDI evaluation included global value of systolic strain and strain rate (Sε, SSR), early diastolic strain and strain rate (Eε, ESR) and late diastolic strain and strain rate (Aε, ASR) were calculated as a mean value for medial segments of septal, lateral, inferior and anterior left ventricular walls. Also ESR/ASR ratio and Eε/Aε ratio were analysed. Correlations between SPL and following TDI parameters: S´, E´/A´, SSR, ESR/ASR, Sε and Eε/Aε were analysed using Pearson correlation coefficient.
Values of Pearson correlation coefficient (r) between SPL and TDI parameters are presented in the table:

	S´	E´/A´	SSR	ESR/ASR	Sε	Eε/Aε
r	0.32	−0.36	−0.68	−0.26	−0.62	−0.49
p	<0.05	<0.05	<0.05	<0.05	<0.05	<0.05

Conclusions: 1. In children after the successful surgical repair of aortic coarctation increased systolic pressure load may suggest affected left ventricular diastolic function and/or elevated left ventricular systolic function. 2. Patient with good results of surgical treatment of aortic coarctation and increased systolic pressure load in ambulatory blood pressure monitoring should undergo periodic echocardiographic assessment of left ventricular function.

P-31

Changes in left ventricular dimension during refeeding in anorexia nervosa associated with increased insulin-like growth factor 1 (IGF-1)

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Patients with anorexia nervosa (AN) are GH resistant, with elevated GH levels and decreased total serum IGF-1 levels. In animal models investigations showed that IGF-1 modulates cardiovascular function and myocardial apoptosis in heart failure. In patients with acromegaly hypertension or LV hypertrophy can be found.

Objective: To investigate how IGF-1 factor modulates LVM and corrected LVM/height 2.7 in the acute state and after refeeding.

Patients and methods: 23 anorexic girls (11.7–17.2 year) who fulfilled the DMS-IV criteria for AN were evaluated at admission and after 9 months refeeding (mean weight gain 0.5 kg/week till minimal healthy weight). Before and at 9 months refeeding all subjects underwent a 2-D-echocardiography (Vivid 7). LVM was calculated according to the Penn Convention. To account linear growth all the LVM data were corrected LVMC by the method of de Simone (LVM divided by indexed *height-indexed to the 2.7 power).

Results:

Parameters	Admission	9 months	p-value
Number	23	23	
Age (y)	13.8 ± 1.54	14.6 ± 1.59	0.097
BMI (kg/m²)	14.6 ± 1.59	17.2 ± 2.19	<0.001
LVM (g)	78.7 ± 20.9	100.0 ± 20.6	0.001
LVM/height (g/m2.7)	27.5 ± 6.64	34.0 ± 5.30	0.001
IGF-1 (ng/ml)	103 ± 58.9	210 ± 78.2	<0.001

Conclusions: We demonstrated that lower levels of IGF-1 were associated with lower LVM and LVMC in the acute state of the disease. The normalisation of the LVM and LVMC was associated with a nearly healthy BMI (17.2 ± 2.19 kg/m²) and IGF-1 level in our patient group. This suggest that IGF-1 is a good endocrine parameter for follow-up of the cardiac mass in AN.

P-32
The role of exercise test in children after correction of congenital heart disease

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The aim of the study was to determine the role of treadmill test /t.t./in children after correction of congenital heart malformations.

Retrospective study estimated t.t. results in 321 children aged 6–18 years (average 11.6 ± 3.5) referred to Cardiology Department between 1.01.2002-31.12.2006, 1 to 16 years after treatment for congenital heart malformations with satisfactory result. Prior to t.t. 57% of children actively participated in sport activities. According the Bruce protocol endurance time, metabolic equivalent, heart rate and blood pressure responses and ECG were analyzed. In 46% patients t.t. were repeated 2–3 times during the period of 6 to 12 months.

Results: Table contains the data concerning the particular indications and abnormal results of t. t.

	TOF after surgery	ASD after interventional treatment	ASD after surgery	VSD after surgery	CoA after surgery	Complex cardiac malformations after surgery	Total
Number of children	61	36	69	76	57	22	321
Abnormal results of t.t	26	5	14	26	17	7	95

Abnormal results of t.t. were due to: exercise induced arrhythmia /39/, exercise systolic hypertension /20/, impaired heart response /33/, and ST-T changes /3/. In group with exercise induced arrhythmia longer follow-up and residual hemodynamic changes were considered as a factors predisposing to arrhythmia. 30% of patients had no arrhythmia detected in 24 hours ECG Holter monitoring. Impaired heart response during t.t. was observed in older patients after TOF, VSD and ASD surgery. Exercise induced hypertension in children operated for CoA in infancy was less frequently observed than in patients older at the time at the surgery (2/22 vs. 15/35).

Conclusion: Despite the good results of treatment in children with congenital heart disease 30% of them reveal abnormalities during t.t.

Children with abnormal result of t.t. should be withhold from maximal physical activity and sport competitions.

P-33
Changes in heart rate and blood pressure with refeeding in anorexia nervosa compared with endocrine parameters

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Objective: To evaluate changes in heart rate and blood pressure during refeeding in patients with anorexia; to identify parameters of endocrine function associated with these differences.

Patients and methods: 23 anorexic girls (11.7–17.2 year) who fulfilled the DMS-IV criteria for AN were evaluated before and 9 months after refeeding (mean weight gain 0.5 kg/week until minimal healthy weight). Biometry, heart rate, blood pressure, endocrine function including IGF-1, TSH, FT4 and 24-h urinary excretion of adrenaline, noradrenaline and dopamine were measured.

Results:

Parameters	Admission	9 months	p-value
Number	23	23	
Age (y)	13.8 ± 1.54	14.6 ± 1.59	0.097
BMI (kg/m²)	14.6 ± 1.59	17.2 ± 2.19	<0.001
HR (bpm)	62.1 ± 21.5	72.3 ± 10.1	0.044
SBP (mmHg)	97.0 ± 13.6	106 ± 8.69	0.013
DBP (mmHg)	58.4 ± 11.6	62.9 ± 4.46	0.089
IGF-1 (ng/ml)	103 ± 58.9	210 ± 78.2	<0.001
TSH (μU/ml)	1.68 ± 1.49	1.14 ± 0.83	0.142
FT4 (pmol/L)	10.1 ± 1.86	11.0 ± 1.83	0.104
24-h urine			
Adrenaline (μg/24h)	2.7 ± 2.68	4.31 ± 3.59	0.132
Noradrenaline (μg/24h)	14.1 ± 9.19	29.2 ± 18.9	0.004
Dopamine (μg/24h)	127 ± 85.4	263 ± 128	0.001

Conclusions: Heart rate and systolic blood pressure significantly rose after refeeding ($p < 0.05$). Heart rate and SBP were inversely related to noradrenaline at admission and after refeeding ($r = -1$). Urine dopamine and serum IGF-1 increased significantly after refeeding. Sympathetic nervous activity reflected the degree of cachexia-underfeeding.

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P-34

Long term postoperative results in isolated discrete subaortic stenosis

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Long-term postoperative results in discrete subaortic stenosis (SAS) are complicated by restenosis (RS), aortic valve injury, reoperation. Factors determining postoperative course are not well defined.
Objective: To assess the influence of anatomic, haemodynamic and surgical factors on the postoperative course in isolated SAS.
Methods: Sixty-six patients operated on at mean age 10.8 ± 3.9 years were studied, 50 membrane, 16 fibromuscular shelf, followed-up 9.9 ± 5.7 years. Resection was performed in 30, enucleation in 36, additional myectomy in 42 patients.
Criteria for unsatisfactory result were NYHA III-IV class, peak gradient (PG) over 50 mmHg, moderate-severe aortic regurgitation (AR) and complete AV block.
RS was defined as PG over 25 mmHg.
Results: There was no early operative mortality. Immediately after operation PG decreased from 85 ± 29 (40–200) mmHg to 13 ± 14 (0–85) mmHg ($p < 0.0001$). Fourteen patients (21%) had PG over 25 mmHg. Three patients (4.5%) had unsatisfactory result – significant PG in 2 and complete AV block in 1.
During follow-up there was 1 late death (mortality 1.5%). RS developed 20 patients (30%). Late result was unsatisfactory in 18 patients (27.3%) due to PG in 16, significant aortic valve lesion in 13 (7 of them with moderate AR). Reoperation was performed in 11 (16.7%). Independent variables predicting unsatisfactory postoperative result were age at operation, immediate PG, narrow aortic ring and annual rate of increase of PG. No one of these variables enter the multiple logistic regression model.
Survival analysis shows freedom from unsatisfactory result in 86%, 71%, 61%, 50.5% at 5, 10, 15 and 18 postoperative year. Independent variables predicting unsatisfactory result were age at operation below 7 years, immediate PG over 25 mmHg and slow rate of aortic annulus growth. No one of these variables enter Cox regression model.
Conclusions: The relatively high rate of unsatisfactory long-term postoperative results is not dependent on the type of SAS and operative technique. The surgery does not interfere the natural course of the disease. No predicting model can be made for the postoperative course in isolated discrete SAS.

P-35

Sexual behaviour and erectile dysfunction in male patients with congenital heart disease

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Objective: Sexuality is an important component of quality of life and subjective well being. Common risk factors for an erectile dysfunction (ED) are chronic illness, especially in heart failure caused by acquired cardiac disease, drugs commonly used in cardiology, and life style. However, there are only little data on patients with congenital heart disease (CHD).
Methods and results: A questionnaire was given to 347 males with CHD (age: 18–69 years; median 23 years) in a specialized tertiary care centre for adults with CHD. Besides various other components the International Index of Erectile Function (IIEF), a generic health related quality of life instrument (SF-12), a depression scale (ADS), and an anxiety inventory (STAI) was included.
6.0% (95% CI 2.7–9.3) of the patients showed signs of ED. There was no significant correlation with the cardiac diagnosis or functional class of the patients. There was also no correlation to known risk factors like age, drugs, alcohol consumption, smoking, and body mass index ($p > 0.05$ each). However, there was a correlation with the psychological sum scale of the quality of life instrument SF-12 and with the depression scale STAI.
Only few patients reported on symptoms during sexual activity like dyspnoea (8.9%), cardiac pain (5.2%) or arrhythmia (8.9%).
Conclusions: This first major study on sexuality and ED in males with CDH shows that ED is also of significance for patients with CHD. Fortunately only few of these to some extend severely limited patients report grave sexual problems. Concerning the correlation between ED and the psychological scales it is not possible to judge from this study, whether psychological issues are the cause or the result of ED. In any case, knowledge of this set of problems is outstanding for caregivers, particularly in specialized centres for adults with CDH.

P-36

Functional outcome in 9-13 year old children after surgical correction in the first year of life because of congenital heart defects

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Introduction: Good functional outcome after total corrective surgery for CHD is generally expected, when performed in the first year of life, but data in literature are scarce.
This study investigates functional outcome in children who had corrective surgery for Tetralogy of Fallot (4F), transposition of the great arteries (TGA) and ventricular septal defect (VSD), all with cardiopulmonary bypass. In addition a group of children with univentricular atrioventricular connection that underwent palliative surgery in the first year and thereafter total cavopulmonary-connection later in life. Functional outcome of CHD children was compared with children in the general population without heart disease.
Method: Parents of 129 survivors of consecutive 4F (31), TGA (29), VSD (50) and TCPC operations (19) in a third referral centre, between 1992 and 1996, were asked to complete the Child Health Questionnaire, parent form. Children with known syndromes were excluded.
Results: There was 81% response (18 TCPC, 23 TGA, 20 4F, 39 VSD). Compared to the general population, the physical summary score (PhS) of 94% TCPC, 64% 4F, 43% TGA and 51% VSD was below p25 (PhS below p5: 53% TCPC, 18% 4F, 22% TGA, 19% VSD). The psychosocial summary score (PsS) of 35% TCPC, 32% 4F, 39% TGA, 49% VSD were below the p25 score of the general population (PsS below p5: 18% TCPC, 14% 4F, 13% TGA, 16% VSD).
Compared to the general population, all four groups scored worse on the physical summary and psychosocial summary. The TGA, 4F and TCPC group together scored significantly lower on the PhS compared to the VSD group (Mann-Whitney, $p = 0.021$); but not on the PsS. Comparing the 4 groups separately with each other revealed a significantly lower PhS of the TCPC compared to the VSD ($p = 0.000$), 4F ($p = 0.007$) and TGA group ($p = 0.002$).

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Conclusion: Many children who underwent corrective heart surgery because of a congential heart defect show a diminished health status 8–13 years after operation, compared to the general population. Physical functioning was most impaired in the Fontan group.

P-37

Maximal aerobic capacity in children operated for congenital heart disease

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Objectives: To assess maximal aerobic capacity (VO_2peak) in children with completed surgical treatment for congenital heart disease (CHD) and in healthy controls.
Material and Methods: VO_2peak was assessed in 42 children, 9–11 years old, with completed surgical treatment of CHD and in 92 healthy age-matched, randomly selected controls. Patients represented a cross section of all patients operated for CHD at our institution. Patients in whom further surgery was anticipated within a year were excluded. Also, patients with chromosomal aberrations and/or neurological dysfunction were excluded. All children performed a ramp test until exhaustion on a cycle-ergometer with measured gas exchange (breath-by-breath (Vmax Spectra 29, SensorMedics, USA)).
Results: VO_2peak was reached in 38 patients and 86 controls, and the results are presented in Table 1. Boys had a significantly higher VO_2peak ($ml \cdot min^{-1} \cdot kg^{-1}$) than girls in both patients ($P < 0.001$) and in healthy controls ($P < 0.001$). Even if there was a significant difference in VO_2peak ($l \cdot min^{-1}$) between patient girls and healthy girls ($P < 0.05$), no difference in VO2peak was seen when controlling for body weight ($ml \cdot min^{-1} \cdot kg^{-1}$). Among boys VO_2peak was similar between patients and controls.

Table1. Physical characteristics and VO_2peak (mean (sd)). T-test for independent groups was used in the comparisons between patients and controls, and between boys and girls.

	Girls		Boys	
	Patient (n = 18)	Control (n = 46)	Patient (n = 20)	Control (n = 40)
Age (yr)	9.8 (0.6)	10.0 (0.5)	9.8 (0.7)	9.9 (0.5)
Weight (kg)	33.2 (6.6)	35.7 (5.6)	34.1 (6.7)	35.7 (6.3)
Height (m)	1.40 (0.07)	1.43 (0.06)	1.40 (0.09)	1.43 (0.07)
BMI ($kg \cdot m^{-2}$)	16.7 (2.1)	17.3 (2.1)	17.2 (2.5)	17.4 (2.7)
VO_2 ($l \cdot min^{-1}$)	1.33 (0.26)	1.51 (0.27)⋆	1.62 (0.25)††	1.73 (0.29)†††
VO_2 ($ml \cdot min^{-1} \cdot kg^{-1}$)	41 (7)	43 (6)	48 (6)†††	49 (8)†††

⋆$P < 0.05$, case vs control ††$P < 0.01$ †††$P < 0.001$, boys vs girls.

Conclusions: In this case-control study of prepubertal children with completed surgical treatment for CHD, exercise performance was within normal limits.

P-38

Outcome after aortic valve replacement with mechanical prosthesis in children and adolescents

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Introduction: Aortic valve replacement using a mechanical prosthesis represents one of the therapeutic options currently available for children presenting with a severe dysplastic aortic valve not suitable for valve sparing surgical procedures. We aimed to study long-term outcome in these patients.
Methods: In this monocentric descriptive study we analyzed clinical records of all patients who underwent surgery for aortic valve replacement with mechanical prosthesis in the time period from January 1992 to December 2005. Excluded were patients presenting with left ventricular outflow tract obstruction requiring an aorto-ventriculoplasty (Konno–procedure). A prospective clinical follow-up examination was performed, including conventional and tissue Doppler echocardiography and magnetic resonance imaging (MRI).
Results: 30 patients underwent aortic valve replacement using mechanical prosthesis (St. Jude Medical, diameter: 23 [17–27] mm (median [range]) at the age of 14.3 [7.6–24.3] years. Indications were severe aortic stenosis in 5/30, aortic regurgitation in 20/3 or a combination of aortic stenosis and regurgitation (4/30). Chronic anticoagulation was established with phenprocoumon at INR 2.5 to 3.5. Follow-up was 6 [1.2−14.5] years. 29/30 patients were in functional class NYHA I. Re-replacement of mechanical prosthesis was necessary in 2/30 patients: in one patient due to somatic growth and in one 31 year old woman with known heterozygous prothrombin mutation (and the intention to become pregnant) due to valve thrombosis while on low-molecular-weight heparin. None of the patients suffered from major bleeding or cerebrovascular accidents during follow-up. On MRI, left-ventricular muscle mass/m^2 body surface was 55 [34–75] g/m^2 and did not exceed the upper limits of controls as obtained in adults by K. Alfakih (J Magn Reson Imaging 2003;17:223–229). Left ventricular ejection fraction (biplane, Simpson) obtained by echocardiography was 55 [24–67]% and showed a good correlation to MRI. Peak strain of left ventricular myocardium (laterobasal) was −13.3 [−27.7 to −0.5]%.
Conclusion: Aortic valve replacement with mechanical prosthesis offers a good option for children and adolescents presenting with severe dysplasia of the aortic valve leading to normalization of left ventricular size and function in most patients.

P-39

Centralized archiving of digital echocardiographic data for mid and long term multicenter studies within the competence net for congenital heart disease in Germany

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Introduction: Clinically approved new echocardiographic parameters including tissue Doppler and 2D strain data for the quantification of regional and global right ventricular function and their long-time follow-up in a large number of patients are

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lacking. Furthermore, the controlled analysis and digital storage for new statistical testing is not yet established.
Setting: Assessment of regional and global ventricular function in controlled prospective clinical studies by the competence net for congenital heart diseases (CNCHD).
Methods: Included in one of the CNCHD studies standardized data sets of conventional and tissue Doppler echo are obtained digitally from all study patients. Therefore a central imaging data management system has been set up. Retrieving the pseudonymized DICOM-Images from the central imaging database the data analysis can take place in three different centers. The results of the analyses are stored in a central Remote Data Entry System (RDE) using the patient's Person Identifier (PID). The PID – a pseudonym automatically generated from the patient's identification data (IDAT) – is the primary key for the medical data (MDAT) in the RDE system as well as for the DICOM-Images in the image database.
Results: Due to the vendor's interpretation of the DICOM standard for Echo, data management of these data still is a challenge. Focusing on a homogeneous set of modalities, echocardiographic data for cross-sectional and longitudinal studies are available for clinical multi-site studies.
Conclusions: By standardized analysis of pseudonymized echocardiographic parameters of the patients included in the CNCHD-studies a quality level for preserving longitudinal data is achieved. These data sets can be used for multicenter studies for evaluating the long term outcome and effects of therapeutic interventions in different congenital heart disease.

P-40

Adult patients after total correction of coarctation of aorta – evaluation of systolic and diastolic function of left ventricle by tissue dopler imaging

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Introduction: After surgical correction of coarctation of aorta (CoAo) patients (P) may still suffer from arterial hypertension (HA) and early manifestation of angina pectoris that may lead to dysfunction of left ventricle (LV).
The aim of the study was evaluation of systolic and diastolic function of LV in adults after correction of CoAo according present HA, residual stenosis of descending aorta and LV mass index using tissue dopler imaging (TDI).
Material and methods: Study group consists of 74 P (35M) aged 19–61 years (mean 31.2 ± 9.8), operated at age 0.5–34 years (mean 10.4 ± 6.8). All P were in NYHA I. Controls: 30 volunteers (18M) aged 26–46 years (mean 32.2 ± 6.6). Following echocardiographic parameters were evaluated: ejection fraction by Simpson (EF), LV mass divided by body surface area – index mass (LWMI), E wave, A wave, E/A, DCT, IVRT, residual stenosis of descending aorta (AoD). In TDI: maximal myocardial velocity of systolic (S´) and diastolic (E´, A´) basal segments of: intraventricular septum (IVS), posterior (PW), lateral (LW), inferior (IW) and anterior wall (AW) were measured. Group without arterial hypertension (HT-)-42P and with arterial hypertension (HT+) 32P, group with larger (LVMI+) 40P, and smaller (LVMI-) 34 P ($M > 103.76$ g/m^2; $F > 99.15$ g/m^2) with residual stenosis (in descending aorta ≥25 mmHg) (AoD+) 32 P and without residual stenosis (AoD-) 41 P were evaluated.
Results: Comparison of classic systolic and diastolic parameters between study group and controls do not demonstrate significant differences. Significant differences were present in TDI between HT- and HT+: S´PW (6.50 ± 2.08 vs 6.70 ± 2.71 $p = 0.02$), S´LW (9.77 ± 3.39 vs 8.70 ± 3.74 $p = 0.04$), S´AW (8.85 ± 2.96 vs 8.20 ± 2.30 $p = 0.03$), Mean S´ (8.23 ± 8.16 ± 1.89 $p = 0.04$), and between LVMI+ and LVMI−: (S´PW (6.67 ± 2.43 vs 5.48 ± 1.40 $p = 0.01$), S´LW (10.30 ± 3.35 vs 7.57 ± 2.39 $p = 0.0003$), S´AW (9.07 ± 2.92 vs 7.11 ± 2.08 $p = 0.002$), Sr. S´ (8.57 ± 1.84 vs 7.21 ± 1.31 $p = 0.0009$). There were no differences in S´ between subgroups AoD+ and AoD- and all measured TDI diastolic parameters in all subgroups.
Results: 1. Systolic and diastolic function of LV in conventional echocardiography in adult patient after total correction of CoAo is preserved.
2. TDI demonstrate dysfunction of systolic function in group of patients with HA+ and larger LVMI+ and preserved systolic function in patients with AoD+.

P-41

Evaluation of Right Heart Myocardial Performance in Patients with Ebstein's Anomaly – a TDI Study

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Introduction: In patients with Ebstein's anomaly non-invasive assessment of right heart function is difficult. We used Tissue-Doppler-Imaging (TDI) echocardiography for non-geometric evaluation of regional and global right heart myocardial performance. Furthermore we analysed the correlation of patient's exercise capacity and right cardiac function.
Methods: 25 patients with native Ebstein's anomaly (mean age: 26.92 years, range 1 to 61 years) and 29 age-matched healthy individuals were examined using TDI. For global performance velocity profiles were measured at the right ventricular (RV) tricuspid valve annulus. Evaluating regional systolic function longitudinal strain was derived at a basal and apical segment of the RV lateral wall, as well as at corresponding regions of the basal (atrialized) and apical (above tricuspid valve closure) interventricular septum (IVS). Strain rate A-wave representing atrial performance was derived at the right lateral atrial wall. For the control group we used analogous segments. Patients underwent exercise-testing. Physical capacity (W/kg), slope and VO^2max were correlated with echocardiographic data.
Results: While RV velocity profiles remained alike in the two groups in systole and early diastole, late diastolic velocity was significantly faster in the Ebstein's group (10.2 ± 4.4 cm/s vs. 7.0 ± 2.8 cm/s; $p < 0.01$). Patients displayed a significant diminution of strain in the basal levels of the RV (25.2 ± 10.5% vs. 36.5 ± 6.9%; $p < 0.001$) and IVS (20.2 ± 6.9% vs. 27.8 ± 7.5%; $p < 0.01$). No significant difference was found in the apical segments. Likewise comparison of right atrial strain rate A-wave remained without significant difference. Mean exercise-capacity was reduced (1.47 ± 0.45 W/kg); mean slope was 36 ± 13 and mean VO_2max was reduced to 20.30 ± 5.95 ml/min/kg. There was no correlation between cardiopulmonary capacity and TDI data.
Conclusion: In patients with Ebstein's anomaly a distinct impairment of regional systolic myocardial contraction can be quantified in basal RV and IVS segments. Nevertheless these segments may still contribute to ventricular ejection. Unaltered global velocities in combination with non-correlation of exercise-testing imply that performance of atrial and apical regions of the RV and IVS are

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able compensate, to some degree, for regional impairment. This is leading to our assumption that right cardiac function cannot account alone for patient's reduced physical capacity.

P-42

Six-minute walk test with measuring wheel: reference values from 3 to 18 years of age

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Objective: To evaluate the 6-minute walking distance for healthy caucasian children and adolescents of a population based sample from the age of 3 to 18 years.
Study design: 280 boys and 248 girls completed a modified test, using a measuring wheel as incentive device.
Results: Median 6-minute walking distance increased from the age of 3 to 11 years in boys and girls alike and increased further with increasing age in boys (from 667.3m to 727.6m), whereas it essentially plateaued in girls (655.8m to 660.9m). After adjusting for age, height ($p = 0.001$ in boys and $p < 0.001$ in girls) remained independently correlated with the 6-minute walking distance. In the best fitting and most efficient linear and quadratic regression models, the variables age and height explained about 49% of the variability of the 6-minute walking distance in boys and 50% in girls.
Conclusion: This modified 6-minute walk test proved to be safe, easy to perform and highly acceptable to children. It provides a simple and inexpensive means to measure functional exercise capacity in children, even of young age, and might be of value when conducting comparable studies.

P-43

Clinical presentation and echocardiographic features of Double Orifice Mitral Valve

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Introduction: Double orifice mitral valve (DOMV) is a rare congenital malformation characterized by the presence of two orifices in the left atrio-ventricular valve area, each having an independent chordal attachment to papillary muscle.
Methods: The retrospective study of 10 patients (pts) who have been examined in our echo laboratory between 1993 and 2006 and in whom DOMV was diagnosed (7 female, mean age 39.3, range 8–59.3 male mean age 28.6 range 15–54) was carried out. The pts were referred to examination with the initial diagnosis of mitral valve disease or congenital heart disease with left-to right shunt. TTE was performed in all cases and 2 of them had additional TEE. 2D images of the parasternal long- and short-axis views, apical 4- and 2-chamber views were inspected for the location and the size of the orifices. Using the different planes of the short-axis view (from the leaflet edges all the way through the valve ring, at the leaflet edge level, at the mid-leaflet level) the type and morphology of DOMV was defined. The study was completed by the evaluation of the systolic pressure gradient across the mitral valve and by assessing the degree of MR.
Results: In 7 pts the complete bridge type of DOMV was recognized, the other 2 pts had the duplicate mitral valve and 1 had the hole type. In all cases mild to moderate mitral regurgitation was present. The mitral stenosis, moderate to severe, has been diagnosed in 6 cases and was associated with the complete bridge type. In 4 pts the ostium primum septal defect was present (1 pt-complete brigde, 2-duplicate MV, 1-hole type). The TTE was sufficient to define the type, anatomy and associated lesion in 8 cases. 2 pts required TEE to confirm the diagnosis because the TTE was equivocal.4 pts with PAVC were operated on.
Conclusions: There are no typical clinical findings attributed to DOMV. TTE especially in short axis parasternal views is reliable method and in most cases sufficient to establish the diagnosis of the DOMV and to determine its type.

P-44

The geometry of the pulmonary artery bifurcation in children – an MRI in vivo study

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Introduction: Reconstruction of the pulmonary artery bifurcation is often required during surgical repair of congenital heart disease. Anomalies of the pulmonary arteries (Pas) may persist after surgery and require reintervention. Information about normal values of the PAs size and three-dimensional geometry may be helpful for planning interventions, but does not exist.
We sought to establish normal values of the size and geometry of the central PAs in children.
Methods: 75 children with normal cardiovascular anatomy underwent contrast-enhanced magnetic resonance angiography (MRA) for investigation of central line related complications. Median age was 10 years (range 2–20), weight 34 kg (10–82), heart rate 80 bpm (59–110). MRA were performed with breathhold in 50 patients and during free breathing in 25. The pulmonary vascular anatomy was reconstructed using subvolume maximum intensity projection (MIP) images in an axial plane, coronal oblique planes along the right and left pulmonary arteries (RPA and LPA), and in a sagittal oblique plane along the main pulmonary artery (MPA).
Results: Image quality was excellent in 31 cases, good in 30, fair in 8 and poor in 3 (excluded from the study).
The MPA shows an oblique course from left anterior inferior to right posterior superior, with an angle of $17 \pm 5°$ in respect to the sagittal plane and an inclination of $57 \pm 8°$.
The MPA bifurcates with a total angle of $99 \pm 10°$; the RPA takes off with an angle $47 \pm 7°$ to the right and the LPA with an angle of $51 \pm 8°$ to the left in respect to the long axis of the MPA. The RPA is caudally angulated by $8 \pm 6°$ and the LPA by $2 \pm 8°$. The origin of the LPA is located 7 ± 2 mm higher than the origin of the RPA. The distance from the bifurcation to the take off of the first segmental branch is 17 ± 6 mm/m^2 for the RPA and 15 ± 5 mm/m^2 for the LPA. The Pas size is listed in table 1.

Table 1. Diameters obtained in two dimensions, normalized to body surface area (mm/m^2)

Vessel	Location of measurement	Diameters
MPA	mid portion	$18 \pm 5 \times 17 \pm 4$
RPA	at origin	$11 \pm 3 \times 13 \pm 3$
	before 1st branching	$10 \pm 3 \times 13 \pm 3$
LPA	at origin	$13 \pm 3 \times 14 \pm 3$
	before 1st branching	$11 \pm 2 \times 11 \pm 2$

Conclusions: The normal geometry and dimensions of the central Pas and their bifurcation in children are described. These data

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may be helpful for indication and for planning catheter-guided or surgical interventions .

P-45

Tissue Doppler Imaging of infantile Pompe Patients shows regional systolic and diastolic Changes during long term Follow Up under Enzyme Replacement Therapy

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Glycogenosis II known as Pompe disease is a deficiency of the glycolytic lysosomal enzyme acid alpha-glucosidase (GAA) with autosomal-recessive inheritance. Generalized myopathy and cardiomyopathy characterize the infantile form, leading to death in early infancy, mostly of cardiac failure. An Enzyme Replacement Therapy (ERT) has been developed and approved for treatment in 2006. Patients receive two-weekly infusions of rhGAA.

We report the follow up of two infants over 16 and 21 months of treatment by now. Both patients presented with neonatal hypertrophic cardiomyopathy and were diagnosed with Glycogenosis II. They were enrolled in an ERT treatment at the age of 2 and 4 months respectively.

Echocardiographic studies including Tissue Doppler Imaging (TDI) were performed before the initiation of therapy and during monthly follow up exams under therapy.

The Echocardiographic images showed a change from hypertrophy to normal of the interventricular septum and the left posterior wall with normalization after 6 and 8 months of treatment respectively. Surprisingly, the shortening fraction showed no changes.

TDI data showed regional increases in systolic and diastolic velocities clearly before normalization of wall diameters and, even more interesting, continuing after.

While the interventricular septum, the left inferior wall and the right ventricle showed early systolic and diastolic improvement with doubling of the velocities over the first months, the left lateral wall showed initially no distinct changes, but started improving after about 6 months of ERT.

While the systolic velocities of the septum and the right ventricle came to a steady state, the diastolic parameters continued to improve.

To our knowledge, this is the first longitudinal Echocardiographic and TDI examination of Pompe patients under ERT.

Tissue Doppler imaging can provide information about systolic and particularly diastolic function on a more sensitive level than conventional Echocardiography. We would therefore suggest a detailed Echocardiographic work-up of Pompe patients including Tissue Doppler imaging.

P-46

Velocity vector imaging analysis of cardiac function in normal neonates

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Objective: Velocity vector imaging (VVI) is a novel angle-independent two-dimensional imaging technique that quantifies the dynamics of cardiac motion. The aim of this study was to define the normal values for cardiac motion and deformation in neonates using VVI.

Methods: Data were collected from 15 healthy term neonates (8 females, 7 males), mean age 1.8 ± 1.01 days, mean birth weight 3.59 ± 0.32 kg and mean length 51.2 ± 2.05 cm. The acquisitions were done in supine position at rest. The apical four chamber view of the heart was analyzed offline using the Axius TM VVI software (Siemens Medical Solutions), for the assessment of longitudinal myocardial velocities, strain and strain rate of the anterolateral wall of the left ventricle, the lateral wall of the right ventricle and the interventricular septum. 2–3 cardiac cycles were averaged. Measurements were performed at basal mid and apical level for each wall. Mean values ± standard deviation were calculated for each wall segment.

Results:

	RVLW		
	MV		
	S	ED	LD
B	4.33 ± 1.61	−2.50 ± 1.49	−4.25 ± 2.19
M	3.05 ± 1.23	−2.20 ± 1.11	−2.88 ± 1.56
A	1.92 ± 1.039	−1.29 ± 1.48	−1.64 ± 1.21
	LVALW		
	MV		
	S	ED	LD
B	3.08 ± 0.98	−3.58 ± 1.60	−2.60 ± 1.22
M	2.25 ± 0.97	−2.47 ± 1.07	−1.88 ± 0.85
A	1.14 ± 0.54	−1.20 ± 0.87	−0.83 ± 0.73
	IVSW		
	MV		
	S	ED	LD
B	3.00 ± 0.78	−2.40 ± 1.05	−2.60 ± 1.53
M	1.98 ± 0.99	−1.81 ± 0.84	−1.95 ± 0.57
A	0.94 ± 0.73	−0.92 ± 0.57	−0.85 ± 0.43

	RVLW			
	MSR			MS
	S	ED	LD	S
B	−1.51 ± 0.82	1.54 ± 1.16	1.25 ± 0.65	−13.49 ± 7.42
M	−1.17 ± 0.40	0.99 ± 0.53	1.00 ± 0.57	−10.41 ± 4.70
A	−0.89 ± 0.41	0.86 ± 0.57	0.97 ± 0.97	−9.76 ± 4.9
	LVALW			
	MSR			MS
	S	ED	LD	S
B	−1.33 ± 0.46	1.28 ± 0.67	0.88 ± 1.22	−13.64 ± 7.25
M	−1.71 ± 0.57	1.39 ± 0.75	1.11 ± 0.43	−17.93 ± 7.88
A	−1.70 ± 0.93	2.00 ± 0.97	0.94 ± 0.38	−16.73 ± 6.31
	IVSW			
	MSR			MS
	S	ED	LD	S
B	−1.45 ± 0.41	1.60 ± 1.14	1.14 ± 0.87	−11.62 ± 4.43
M	−1.92 ± 0.69	2.21 ± 0.70	1.12 ± 0.55	−17.78 ± 5.38
A	−1.24 ± 0.92	2.04 ± 0.83	0.75 ± 0.38	−13.49 ± 4.66

Abbreviations: Basal segment: B, Mid level segment: M, Apical segment: A, Right ventricular lateral wall: RVLW, Left ventricular anterolateral wall: LVALW, Interventricular septum wall: IVSW, Myocardial velocity: MV, Myocardial strain: MS, Myocardial strain rate: MSR, Systolic period: S, Early diastolic period: ED, Late diastolic period: LD

Conclusion: This study shows the feasibility of using VVI to quantify myocardial motion and deformation in newborns. This angle-independent method needs further validation, looking at the reproducibility and comparing it to one-dimensional techniques.

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P-47

Assessment of Left Heart Myocardial Performance in Patients with Ebstein's anomaly – a TDI study

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Introduction: Quantification of left heart function using conventional echocardiography in patients with Ebstein's anomaly is challenging. We used Tissue-Doppler-Imaging (TDI) echocardiography for non-geometric quantification of global and regional left atrial and ventricular myocardial performance. Furthermore patient's reduced physical capacity cannot be explained solely by right cardiac function, thus we correlated exercise-testing with echocardiographic left ventricular (LV) data.
Methods: We examined 25 patients with native Ebstein's anomaly (mean age: 26.92 years, range 1 to 61 years) and 29 age-matched healthy individuals using TDI. For global performance velocity profiles were measured at the LV mitral valve annulus. To calculate TDI Tei-index we appraised time spent in isovolumetric relaxation (IVRT), contraction (IVCT) and ventricular ejection (ET) via velocity curved m-mode at the interventricular septum. Evaluating regional systolic function longitudinal strain was derived at a basal, middle and apical level of the left lateral wall, radial strain at the LV posterior wall. Strain rate A-wave representing atrial performance was derived at the left lateral atrial wall. 14 patients underwent bicycle exercise-testing. Physical capacity (W/kg) was correlated with echocardiographic data.
Results: In Ebstein's anomaly LV velocity traces were significantly slower in systole (5.3 ± 2.19 vs. 7.51 ± 1.91 cm/s) and early diastole (7.62 ± 3.13 vs. 11.30 ± 2.52 cm/s; both $p < 0.001$). Patient's Tei-index was significantly higher (0.49 ± 0.11 vs. 0.32 ± 0.04; $p < 0.001$), due to prolongation of IVRT (85.13 ± 15.04 vs. 58.65 ± 14.86 ms; $p < 0.001$) and IVCT (53.05 ± 10.16 vs. 44.60 ± 11.07 ms; $p < 0.01$) and shortening of ejection time (285.89 ± 38.94 vs. 321.55 ± 27.52 ms $p < 0.001$). Furthermore a significant diminution of longitudinal strain (basal: 17.99 ± 5.74 vs. 24.76 ± 8.46%, middle: 15.96 ± 5.27 vs. 21.41 ± 6.23%, apical: 14.53 ± 4.88 vs. 19.22 ± 4.75%; all $p < 0.01$; mean strain: 16.04 ± 4.34 vs. 21.82 ± 5.52%; $p < 0.001$) and radial strain (30.42 ± 19.00 vs. 45.28 ± 11.05%; $p < 0.001$) was found. Atrial strain rate A-wave showed no significant difference. Physical capacity was reduced (mean 1.47 ± 0.45 W/kg) and correlated well with velocity E-wave ($p < 0.05$), IVRT ($p < 0.05$) and mean LV strain ($p < 0.01$).
Conclusions: In Ebstein's anomaly a distinct impairment of global and regional left ventricular myocardial contraction can be quantified, affecting both systole and early diastole. We assume that this accounts for patient's reduced physical capacity. Remarkably their left atrial function is unrestricted.

P-48

Interventricular septum function after early and late TOF correction, a Tissue Doppler Study

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Aim of the Study: To detect the function of the interventricular septum late after repair of Tetralogy of Fallot (TOF), to detect any difference between patients who had undergone correction before and after three months of age and to detect any difference between patients with or without complete right bundle branch block (QRS > 120 msec).
Methods: Doppler pulsed TDI (Tissue Doppler Imaging) of the interventricular septum at the level of the mitral valve anulus were performed in 8 healthy subjects and in 15 patients who had undergone surgery for TOF randomly selected from our database. In the surgical group 5 patients had undergone TOF correction under three months of age and 10 patients after this age. The following TDI parameters were evaluated: systolic peak velocities (Sm), early (E(m)) and late (A(m)) diastolic velocities, E(m)/A(m) ratio and myocardial acceleration during isovolumic contraction (IVA).
Results: Compared with healthy children, Doppler tissue imaging velocities for patients with TOF showed decreased myocardial velocities during early diastole ($P = 0.02$), systole ($P = 0.02$) and decreased IVA ($P = 0.02$). Comparing the two surgical subgroups, patients corrected after 3 months of age showed decreased IVA ($P = 0.0007$), increased myocardial velocities during late diastole ($p = 0.02$) and systole ($p = 0.04$). There were no differences between patients with or without complete right bundle branch block.
Conclusions: Patients late after correction of TOF showed impaired interventricular septum function compared to normal subjects. Our small study seems to demonstrate that to an early correction correspond a better results in terms of function of the interventricular septum.

P-49

Impact, safety and hemodynamic effects of magnetic resonance imaging in pediatric cardiac intensive care patients

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Objectives: MRI has proven to be an important diagnostic tool in congenital heart disease. The use of MRI in critically ill children awaiting cardiac surgery or after open heart procedures has been restrictive in the past due to the challenging MRI environment which carries several inherent risks.
We hypothesized that cardiac MRI examinations can be performed with low risk and without significant hemodynamic consequences in this highly selected patient group.
Methods: Retrospective analysis using electronic records of all patients submitted from our PCICU to our cardiac MRI-program in the last two years.
Data collected included age, diagnosis, inotropic score, urine output, medication, temperature, lactate, length of MRI, adverse effects, clinical implications of MRI and length of stay in the PCICU.
Results: 24/592 patients were referred from our PCICU. 22 infants, 2 schoolkids, 10/24 were postoperative. Intracardiac malformations were present in 19, vascular rings/tracheal stenosis in 3, 1 cardiomyopathy and 1 mycordial infarction. Mean stay in the PCICU was 25.7 days. 8 were ventilated, 6 were on inotropic support. All nonventilated children were intubated for the MRI and extubated in the MRI-laboratory. Mean duration of MRI ("door to door-time") was 108 minutes. All children except one were hemodynamically stabile and no increase of catecholamines was necessary during and after the scans. A 10-month old girl needed a simple dose of epinephrine because of relative bradycardia after a breathhold-sequence. Mean temperature after readmission to the PCICU was 36.9°C, mean serum-lactate 1.2 mmol/l. Mean

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diuresis on examination day was insignificant better than the day before with unchanged medication (603 ml vs. 644 ml). In 18/24 patients operative or catheter-interventional procedures were initiated after the MRI result.
Conclusions: Cardiac MRI is of high clinical value and can be performed with low risk and no negative hemodynamic effects even in pediatric cardiac intensive care patients.

P-50
Comparison of Non-Invasive Cardiac Output Estimation Using Applanation Tonometry and Transmission Line Theory Versus Conventional Doppler
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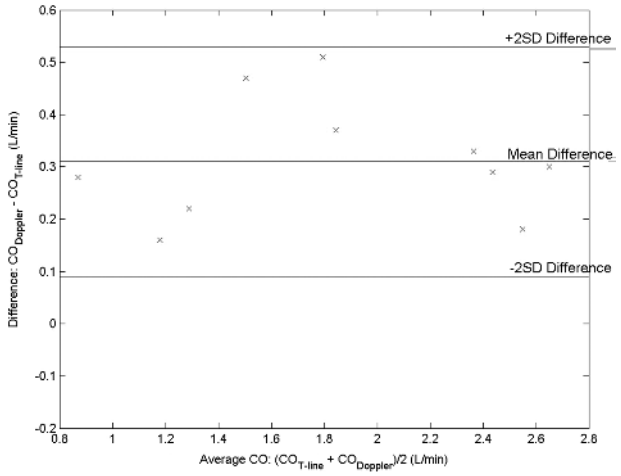
Introduction: Transmission line theory has been proposed as a method of determining cardiac output (CO) using the arterial pulse wave. With this method the arterial system is modeled as a uniform transmission line. For a given arterial pulse wave, the aortic flow can theoretically be calculated from the model.
Purpose: To compare CO measurements determined by applanation tonometry with transthoracic Doppler measurements.
Methods: Ten healthy children with a mean age of 10.1 ± 2.3 years participated in this study. Calibration was done by acquiring the aortic velocity profile using conventional Doppler methods and the carotid pressure tracing using applanation tonometry simultaneously. During calibration, several arterial system parameters were identified by grey-box identification and optimization. The aortic diameter was measured by two-dimensional echocardiography and was used to convert aortic velocity to volumetric flow. Three consecutive cardiac cycles were used during calibration. After 5 to 15 seconds the aortic velocity profile and carotid pressure were simultaneously reacquired. Three consecutive cardiac cycles from the carotid pressure tracing were chosen to calculate the CO using the transmission line method. The results were compared to the CO determined from the velocity-time integral of the Doppler velocity profile. Results: The mean CO determined by the transmission line algorithm and transthoracic Doppler were 1.69 ± 0.63 L/min and 2.00 ± 0.63 L/min, respectively. The mean difference was 0.31 ± 0.11 L/min. The mean percentage difference was 18.8 ± 8.8%. The correlation coefficient (r) was 0.98 ($p < 0.0001$). The Bland-Altman limits of agreement were between 0.09 and 0.53 L/min (95% Confidence Interval). The Bland-Altman plot is shown in Figure 1.



Conclusions: This new method gives consistent non-invasive measures of CO which correlate well with other known methods. Further evaluation is needed to determine whether this technique may be used to monitor continuous, non-invasive CO from the arterial pulse.

P-51
Perioperative Stent Implantation for Management of Pulmonary Artery Stenoses in Infants with Single Ventricle Physiology
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Objectives: To evaluate the efficacy and safety of stent implantation to facilitate perioperative management of pulmonary arterial stenoses in infants with single ventricle physiology.
Patients and Methods: Nine infants (weight range 4.7–9.6 kg; age 3–6.5 months) with complex single ventricle physiology were evaluated by cardiac catheterization immediately following (3–18 days post-surgery) a bidirectional superior cavopulmonary (bidirectional Glenn – BDG) shunt, due to persistent SVC syndrome, hypoxemia, and raised SVC pressure (mean 18–25 mmHg). Single or multiple stenoses affecting the left pulmonary artery (either directly behind a Damus anastomosis (n = 7) or at the BDG anastomosis (n = 2)) or the right pulmonary artery (n = 3, at the BDG anastomosis) were identified. The minimum diameter of the stenoses ranged from 1.5 mm to 3.2 mm. Via the internal jugular vein, premounted Palmaz-Genesis stents (on 6, 8 or 10 mm diameter balloons) were implanted across the stenoses.
Results: All stents were successfully implanted. The final diameter of the treated segment ranged from 5.8 to 9.6 mm. One infant had a tear of the left pulmonary artery, at a site remote from the surgical scar, which required immediate reoperation and re-repair. In the remaining patients, the SVC pressure decreased to between 14–18 mmHg within 72 hours of the procedure. Six patients have undergone successful Fontan completion; 2 are awaiting further evaluation and surgery. One patient (with LPA tear) died as a result of recurrent PA thromboses due to an as yet uncharacterised familial coagulation disorder.
Conclusions: Perioperative stent therapy may be life-saving. It produces effective relief of stenoses without the need for surgical reintervention, and improves the clinical outcome of this subgroup of patients. Stent implantation across suture lines in the immediate post-operative period appears to be safe.

P-52
Cor triatriatum dexter as a contraindication for transcatheter closure of secundum atrial septal defects with the Amplatzer Septal Occluder
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Introduction: Cor triatriatum dexter (CTD) results from persistence of the right valve of the sinus venosus and has varying clinical manifestations, depending on the degree of partitioning or septation of the right atrium. A large membrane dividing the right atrium can cause right-sided heart failure, elevated central venous pressures secondary to obstruction of the tricuspid valve, the right ventricular outflow tract, or inferior vena cava. Often CTD is

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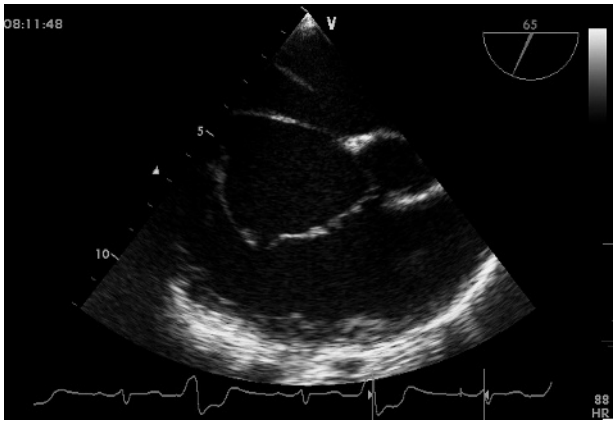
asymptomatic and detected incidentally during echocardiography of associated ASDs.
Methods: 476 pts – 320 female and 156 male, median age 43.2 years (from 1997 to 2007) were accepted for atrial septal defect occlusion, on the basis transthoracic and transesophageal echocardiography (TEE). Transcatheter ASD procedures performed under TEE and fluoroscopic guidance.
Results: 15 pts were operated on for double or multiple defects after control TEE during balloon sizing.
In 11 pts with secundum ASD it was visualized membrane of Eustachian valve joined with the inferior part of the anterior rim of the defect. 7 pts with ASD II and CTD were operated on.
454 transcatheter procedures were performed; in 1 pt with CTD device dropped after releasing to the left ventricular outflow tract and required surgical removal, in 3 pts with CTD it was given up the procedure after TEE during catheterization. Total occlusion was performed in 450 pts with device size from 12 to 40 mm.



Conclusions: CTD as a huge Eustachian valve connected with an anterior rim of the secundum ASD is a morphological contraindication for transcatheter closure with ASO. The persisted membrane of the right valve of the sinus venosus makes it impossible to place the device in a stable position.

P-53
Outcome of Transcatheter Closure of Patent Ductus Arteriosus in Children Using the Amplatzer Duct Occluder

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Introduction: There are no studies reporting late outcome following transcatheter closure (TC) of the patent ductus arteriosus (PDA) using the Amplatzer duct occluder (ADO). In this report we review initial and 5-year results following TC of PDA using the ADO.
Methods: Between April 1999 and May 2001, 126 patients, ages 0.1 to 15 years, with a moderate to large PDA underwent TC using the ADO. All patients had echocardiographic follow-up at 24h, 6 and 12 months after closure and at 12 months intervals thereafter.
Results: The mean PDA diameter was 4.8 mm (range 2 to 11. 5 mm). The mean ADO diameter was 7.2 mm (range 4 to 14 mm). 121/126 patients (96%) had successful device deployment. Complete angiographic closure was observed in 112/121 patients (92.5%). Complete occlusion at one year follow-up was documented in 120/121 patients (99%). 11/121 (9%) patients (<2 years of age-device ≥6–8 mm)) developed a gradient greater than 10 mmHg across the left pulmonary artery immediate or at follow-up. 9/121 (7.5%) patients (<1 year of age-device = 6–8 mm) had an aortic gradient of >10 mmHg immediate or at follow-up. Only 3 patients had an aortic gradient over 20 mmHg. No late evidence of device failure, recanalization, thromboembolism, endocarditis, or hemolysis was observed.
Conclusions: Our late results prove that TC using the ADO can be safely and effectively applied in most patients with a moderate to large PDA. ADO is not an ideal device for closure of certain anatomic type large PDAs in small patients.

P-54
Stent or Balloon for stenotic pulmonary arteries? Short and midterm results

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Introduction: The aim of this study was to compare results of stent implantation and balloon angioplasty (BAP), both established interventional procedures, for the treatment of pulmonary artery stenosis (PAS).
Methods: We reviewed the haemodynamic and angiographic records of 47 PAS patients aged 0–19.5y (median age 2.9y, median BSA 0.59 m^2) who underwent 56 stent implantations and 37 patients aged 0.5–20y (median age 2.8y, median BSA 0.58 m^2) who underwent 49 BAP between 2002 and 2006 at both our institutions. In the BAP group 7 patients had native peripheral pulmonary artery stenosis (PPS), 7 common arterial trunk (TAC), 8 Fontan, and 15 Tetralogy of Fallot (ToF). In the stent group 28 patients had ToF, 3 TAC, 4 PPS, 4 Transposition of Great Arteries, 6 Fontan, 2 homograft. Criteria for interventional approach were: stenosis of arterial lumen ≥40%, right/left ventricle pressure ($P_{RV/LV}$) ratio ≥50% and systolic pressure proximal to the stenotic lesion of the PA ≥25 mmHg. Mann-Whitney testing and analysis of covariance were used for statistical analysis. Statistical significance was set at $p < 0.05$.
Results: The improvement in absolute PA diameter, PA diameter/BSA index and $P_{RV/LV}$ was significantly better in the stent compared to the BAP group (Table).

	BAP (range and median)		Stent (range and median)		
	Pre-BAB	Post-BAB	Pre-stent	Post-stent	P
PAs-diameter (mm)	(1.2–14.4) 6.3	(1.2–15.2) 10.7	(0.9–9.5) 3.4	(3.1–16.8) 7.15	.002
PA-indices (mm^2/m^2)	(3.75–20.17) 8.8	(3.75–22.45) 11.17	(0–19.79) 6.87	(6.15–27.75) 13.75	.006
PRV/LV (mmHg)	(0.16–1.2) 0.56	(0.14–1.12) 0.53	(0.21–1.74) 0.82	(0.07–0.97) 0.60	.022

Complications were 8% in the BAP group (1 PA aneurysm, 1 PA rupture, 1 haemopericardium, 1 balloon rapture necessitating vein cutdown), and 9% in the stent group (2 dislodgments, 1 atrial flutter, 1 broken stent, 1 pulmonary oedema).

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Conclusions: Stent implantation in PAS is a feasible and safe procedure, offers better short and mid-term results than BAP even for small patients. Further redilation of the stents according to patient's body growth will be necessary particularly for infants and young children. Decrease of the pressure overload is significantly better after stent implantation than after BAP but long term benefits remain to be proven.

P-55

Interventional catheterisation of systemic veins: early and long term results

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Background: Vascular stenosis and anomalous vessels prompting right to left shunt are frequent findings in patients with congenital heart disease (CHD) and with other chronic diseases. We reviewed our experience concerning interventions on systemic veins over a 10 year period.
Patients and Methods. From January 1996 to June 2006, 45 patients (34 with and 11 without CHD) underwent catheterisation to occlude collateral vessels (12 with and 4 without CHD) or to dilate stenotic veins (22 with and 7 without CHD). Long term results were evaluated with cardiac catheterisation (n = 10) or CT scan (n = 15).
Results: Median age and weight were 3.7 years (0.1–43) and 39 kg (2.5–87), respectively. 35 stents, 8 occluding devices and 11 coils were implanted. In all but 2 patients it was possible to treat the lesion. There were 2 procedural complications (superior caval vein tear, leading to cardiac tamponade in 2 patients with Senning operation and complete occlusion of the superior caval vein channel). There were no procedural deaths.
At a median follow up of 6 years (1–10), 14 overseas patients were lost. There were 4 late deaths (3 CHD with heart failure and 1 non CHD patient with pulmonary fibrosis). Among 25 patients with complete follow 18 had stent implantations (20 stents) and 7 anomalous vessel embolisation. We observed 4 stent occlusions (20%), 1 stent fracture (5%), 2 repermeabilsation of anomalous vessels (22%). The percentage of stent thrombosis and repermeabilisation of anomalous vessels was similar in patients with and without CHD.
Conclusions: Interventional catheterisation of systemic veins offers good immediate results and a low incidence of complications. However, repermeabilisation of occluded vessels and late stent thrombosis raise concern on long term results.

P-56

Hemodynamic assessment and types of interventions in symptomatic post-Fontan patients

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Some children after Fontan completion clinically deteriorate, requiring hemodynamic assessment. The investigation aimed at assessing hemodynamic evaluation results in post-Fontan children depending on their initial congenital heart defect (CHD) type. The material included 18/170 (10.5%) post-Fontan children (52 – HLHS; 118 – other CHD) operated on in 1989–2006, referred for hemodynamic assessment due to increased fatigability and decreased SO_2 (≤90%). They were divided into two groups: Group 1 – HLHS (N = 7), and Group 2 (N = 11) with other CHD and functionally single ventricle types. The assessment of intergroup differences is presented below.

Group/n	Age at operation (years)	Fontan-cardiac cath interval (years)	SO_2 (%)	mPAP mmHg
1 7/52	2.2 ± 0.3	0.8 ± 0.3	87.6 ± 5.4	17.5 ± 6.2
2 11/118	4.6 ± 3.2	3.1 ± 2.4	87.2 ± 11	12.4 ± 4.1
p = 0.08	p < 0.001	p < 0.002	p = 0.09	p < 0.001

Group/n	trans-pulmonary-pressure D mmHg	SVEDP mmHg	Nakata Index mm^2/m^2	Qp/Qs
1 7/52	7.9 ± 2.2	9.3 ± 5.6	166.6 ± 50	0.75 ± 0.2
2 11/118	3.9 ± 1.6	8.5 ± 3.6	263 ± 77	0.81 ± 0.2
p=0.08	p < 0.002	p = .0.06	p < 0.001	p = 0.08

Group 1 children required diagnostic management significantly earlier, showed higher pulmonary arterial pressure, transpulmonary pressure gradient, and a significantly lower Nakata Index. Interventions in Group 1 and 2 patients included: Group 1 – balloon LPA angioplasty combined with MAPCA closure (4), and azygos vein (av) closure (1); Group 2 patients (fenestration closure – 2, MAPCA closure – 2, interatrial tunnel-common atrium fistula closure – 1, av closure – 1). Two Group 1 patients died of congestive heart failure, 1 was reoperated (interatrial communication enlargement), 2 – disqualified from further treatment. In Group 2, 2 patients died, 2 were disqualified, 2 – reoperated, while in 1, hemodynamic causes of exudative enteropathy were excluded.
Conclusions: In HLHS patients, early disadvantageous circulatory changes are encountered more often.

P-57

National registry for treatment of coarctation of the aorta with stents

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Objectives: The current study was designed to assess the early and midterm results of stent implantation for treatment of native (CoA) or recurrent coarctation (RCoA) of the aorta in children and young adults, on the base of a national registry.
Material and methods: One hundred two stents were implanted in 101pts with CoA (72pts) and RCoa with failed balloon angioplasty (29pts). Median pts age was 16yrs (range 0.01–57yrs). All but one 3day old newborn with duct-dependent CoA with complex congenital heart defect had arterial hypertension. In 4pts stents initially properly implanted migrated to descending aorta shortly after implantation. Due to good haemodynamic effect of dilation with gradient reduced from mean of 53 mmHg to11.75 mmHg, no additional stents were implanted in these pts, who were excluded

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from the analysis. In 12pts with critical CoA, balloon angioplasty with 6mm diameter balloon was performed 3mths before the stent implantation. In 5pts staged stent implantation was decided with elective redilation 3mths later.
Results: Gradient reduction from 41.14+20 mmHg to 5.34+ 6.56 mmHg and increased diameter of aorta from 8.3+2.9 to 15+3 mm was achieved in 94pts. RCoA resistant to stent dilation occured in 3pts, of whom 2pts had had previous dacron patch repair.
Complications: Stroke in a 53yr old pt after covered stent implantation, with no neurological sequele in 8mths follow up, small 3mm diameter aneurysm after stent implantation in 1pt. During mean 35.26+24.6mths follow-up, in 4pts stent fracture was recognized, 3 of whom received a PTFE covered stent due to additional in-stent stenosis and 1 had balloon redilation, in 9pts successful stent redilation for neointimal hyperplasia and/or patient growth 3–6 yrs after implantation were performed. Follow up CT scans 3–6 mths after implantation confirmed good stent position with wide patency. In 56% of pts, antihypertensive treatment was stopped, in 41% reduced, in 3% stent therapy was not sufficient and need surgical reoperation.
Conclusion: Stent implantation for the majority of patients with recurrent and native coarctation of the aorta is safe and effectively reduces the blood pressure gradient across the coarctation site. Arterial hypertension persists in a significant number of the patients.

P-58
Catheterinterventions to reduce frequency of reoperation in patients after repair of common arterial trunc
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Basis: Surgical correction of common arterial trunk (TAC) is usually performed in infancy and often includes implantation of valved graft in pulmonary arterial position. This study evaluates the outcome of grafts and the effect of interventional treatment of grafts and pulmonary arteries in patients after TAC repair.
Methods: 38 patients were reviewed after TAC repair from 1986 to 2005. All patients underwent catheterisation. Interventional treatment with dilatation or stent implantation into the right ventricular outflow tract (RVOT) and pulmonary arteries was performed in order to postpone reoperation.
Results: Median patients age at surgery was 35 days (7days to 9.5years). 34 human grafts and 4 xeno-grafts were implanted with a median size of 12mm (8–20mm). Freedom from reoperation was 97.4% (1 year), 68.4% (5 years) and 47.4% (10 years) respectively. 5 patients needed graft replacement after 11, 12, 15, and 16 years. 13 patients still life with the first graft. Median time from operation in this group is 8.5 years (1.5 to 18.5 years). Patients with reoperation within five years had grafts with a median size of 11 mm (8 to 13 mm). Grafts of patients with reoperation later than 5 years had median size of 13 mm (8 to 19 mm). Indications for reoperation were graft failure and high RV pressure in all but one patient with endocarditis. 27 patients had interventional treatment during catheterisation. 23 patients had 36 catheterisations with dilatation of pulmonary arteries and graft. Four patients had 11 catheterisations with dilatation and implantation of stents. One patient had stent implantation into the right pulmonary artery. The other 3 patients had four stents implanted into the RVOT. Right ventricular pressure could be reduced in all patients with stents and in 13 patients with dilatation.
Conclusion: Outcome of valved grafts after TAC repair is encouraging after ten years. The smaler grafts implanted in infancy tended to be replaced earlier than the larger grafts. High right ventricular pressure due to graft stenosis or pulmonary artery branch stenosis is the most common indications for reoperation. Interventional treatment, particularly stent implantation to RVOT or pulmonary arteries can postpone reoperation.

P-59
Percutaneous Treatment of Conduit and Pulmonary Arteries' stenosis in modified Norwood procedure (Sano modification)
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Introduction: We report on the experience of reliefing stenosis in conduit and Pulmonary Arteries (PA) ussing Cutting Balloon (CB) and stents in patients with Sano procedure.
Method: In the period of time between January 2002 to December 2006 we treated 10 patients (weight 2900–5700 grs.) with Hypoplastic Left Heart Syndome undergone modified Norwood (RV-PA Conduit). Six were presented to the catheterization laboratory for hypoxemia (O2 Sat < 65%), and Four were in ECMO for low cardiac output and/or hypoxemia after surgical intervention; 12 premounted stents (diameter 4–5 mm) were implanted (7 in the Conduit, and 5 in the PA) in 8 patients; 3 CB were used in PA in two infants (one previously to stent).
Results: In 9 patients we obtained a good angiography results (improvement in the diameter stenosis >50%).
The four in ECMO died for cardiac failure but no for hypoxia. Five increased O2 sat > 75% and Bidireccional Glenn were performed before the follow three months. Two children suffered a transiet AV block. In one neonate we produced a disection in the distal conduit anastomosis, a cover stent implantation was needed.
Conclusion: Percutaneous deployment of premounted stents and CB could be a good approach to this kind of patients to relief the hypoxemia delaying the Glenn until the infants get an appropiated weight.

P-60
Transcatheter closure of patent ductus arteriosus among native high-altitude habitants
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Purpose: Patent ductus arteriosus (PDA) is frequently seen in native habitants of high altitudes. We present our experience in transcatheter closure of PDA performed in the highest capital in the world – La Paz (Bolivia). The procedures were done during teaching courses.
Material and method: Ten consecutive patients (pts) with PDA were included in the study. Their age was 14 (1.1–50) year (y), weight 26 (9–66) kg. All of them were citizens of La Paz. In 9 pts type A, in 1 – type E of PDA (according to Kirschenko qualification) were found. Mean pulmonary artery pressure (MPAP) was 34 (7–80) and mean aortic pressure – 77 (58–100) mmHg. Minimal diameter

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of PDA was 4.9 (1.5–8.5) mm and lengths 8 (6–12) mm. One patient – 4.3 y old girl had additionally valvular aortic stenosis (AS) with gradient left ventricle – aorta (LV-Ao) 87 mmHg (in ECHO-Doppler).
Results: In 9 pts (with PDA > 2.5 mm) interventional catheterization with Amplatzer Duct Occluder (ADO) was performed. In 2.4 y old child with PDA of 1.5 mm diameter – detachable coil was implanted. In 2.5 y old girl and 50 y old pt with wide PDA sever pulmonary hypertension (PH) was diagnosed. In both balloon occlusion test (BOT) of PDA showed 50% reduction of MPAP. ADO implantation has failed in the child with PH because the 12/10 ADO went through the 6 mm duct. Subsequent surgery was done without any complications. In case of concomitant AS diagnostic catheterization showed the LV-Ao gradient of 40 mm and aortic valvuloplasty was suspended. Successful closure of PDA with 10/8 ADO produced permanent reduction of this gradient also in ECHO. No residual shunt after 24 hours was observed in any case after successful implantation of ADO.
Conclusions: Patent Ductus Arteriosus in native habitants of highland have different physiopathology and anatomy. Amplatzer Duct Occluder seems to be the ideal devise for transcatheter therapy for most of such patients. It is necessary to develop more interventional cardiology in the countries having such conditions as predominantly cardiosurgery methods exist there.

P-61
Taussig–Bing anomaly – late results after anatomic correction
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Introduction: Taussig–Bing (T–B) anomaly is defined as double outlet ventricle with malposition of the great arteries and subpulmonary VSD. It often coexists with aortic arch anomalies (AAA). Nowadays, a method of choice in treatment of this pathology is an arterial switch with closure of the VSD and aortic arch plasty.
The aim of the study was to evaluate the results of correction of T–B with the estimation of the risk of complications leading to catheter or surgical interventions.
Methods: Between March 1993 and March 2006 41 corrective surgeries of T–B anomaly were performed in our institution. 33% of the patients were neonates (80.5%). There were 31 males (75.6%). In 22 patients (53.7%) AAA was diagnosed: coarctation in 7 patients, coarctation with aortic arch hypoplasia in 12 patients and interrupted aortic arch in 3 patients.
Abnormal origin of the coronary arteries was found in 18 patients (43.9%). Preoperative diagnosis was based on cardiac ECHO, rarely supplemented by angiography.
Preoperatively 29 patients were treated with prostaglandin (70.7%) and 8 patients with balloon atrioseptostomy (19.5%). Postoperative evaluation was based on physical examination, ECG, ECHO and in some cases angiography.
Results: In the group of 41 there were 7 deaths (17.1%) – 6 early and 1 late. Risk factors of death were aortic arch and coronary artery anomalies. There was no statistical difference in the age or the preoperative management of the patients between the group of survivors and the group of those who died. 6 patients (17.6%) needed reoperation – 3 due to RVOTO, 2 due to recoarctation and 1 due LVOTO. In 4 patients (11.76%) catheter interventions were performed: in 2 balloon plasty of recoarctation and in 2 plasty of pulmonary artery stenosis. There was statistical significance in the reoperations in the group with AAA. 7 patients (20.5%) had postoperative pulmonary artery stenosis and 28 patients (82%) trivial/mild aortic valve insufficiency.
Conclusions: Anatomic correction of T–B anomaly is associated with greater mortality, risk of complications and need for surgical or catheter reinterventions compared to simple TGA. The pathology of the aortic arch is the main risk factor for reinterventions and reoperations.

P-62
Myocardial oxygenation and substrate metabolism in pediatric heart surgery: role of cardioplegia
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Introduction: We have previously reported hemodynamic and myocardial metabolic differences related to cardioplegia type in infant heart surgery. The purpose of this study was to clarify the relationship between cardioplegia type and other clinical variables, myocardial oxygenation and metabolic changes after cardioplegia in pediatric heart surgery.
Methods: Seventy-six children, aged four weeks to 5.3 years and undergoing open-heart surgery with cardioplegia (St Thomas' II crystalloid cardioplegia or blood cardioplegia) were included. Oxygen saturation and concentrations of substrates and amino acids were measured in arterial and coronary sinus blood after weaning from bypass.
Results: The uptake/release of alanine ($r = 0.47$, $p < 0.0001$), aspartate ($r = -0.29$, $p = 0.01$), glutamate ($r = -0.48$, $p < 0.0001$), beta-hydroxybutyrate ($r = -0.50$, $p < 0.0001$) and lactate ($r = 0.37$, $p = 0.001$) were related to myocardial oxygenation (estimated as the coronary sinus saturation). In the multivariate analysis, crystalloid cardioplegia, as opposed to blood cardioplegia, and decreased post-operative arterial hemoglobin oxygen saturation were associated with decreased post-operative myocardial oxygenation. Crystalloid cardioplegia and the preoperative medication use for heart failure were associated with greater post-operative myocardial lactate release. A decreased myocardial oxygenation and crystalloid cardioplegia predicted more uptake of glutamate.
Summary: Myocardial metabolic alterations in pediatric heart surgery are related to myocardial oxygenation. The better cardiac function in the period after weaning from bypass after cold blood cardioplegia may be the result of the improved myocardial oxygenation and the reduced need of metabolic adaptive measures.

P-63
Repair of congenital mitral valve anomalies in children: Long-term results in 52 cases
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Introduction: Congenital malformations of the mitral valve (MV) are rare and their surgical management in the pediatric age group remains a therapeutic challenge. We reviewed our experience with repair of MV malformations in the pediatric age group in order to assess early and long-term outcomes in terms of survival and freedom from re-operation.
Methods: Retrospective review of all patients operated between January 1994 and December 2006 for congenital mitral defects.

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Results: 52 consecutive children with congenital MV disease underwent valve repair for: mitral insufficiency (35 patients), mitral stenosis (10 patients) and mixed lesions (7 patients). Associated cardiac anomalies were found in 16 patients (30%). The mean age was 2 ± 1.2 years (2 months–4 years) at the time of diagnosis and 6.9 ± 5years (2 months–14 years) at the time of surgery. Type I (mitral annular dilatation), II (leaflet prolapse) and III (restricted leaflet motion) pathologies were respectively documented in 28, 5 and 17 of the patients. Appropriate surgical techniques were selected according to the pathophysiological mechanisms and a biodegradable ring (Bioring®) was implanted if an annuloplasty was indicated (42 patients). There was no perioperative or long term mortality. Five patients with type III mitral valve pathologies underwent valve replacement: hammock valve (2), short chordae (2) and parachute valve (1).
Conclusions: Our results suggest that reconstructive surgery for congenital mitral valve disease is effective and carry a low risk. Type I and II pathologies have better long term results – probably due to a better learning curve experience – compared to type III lesions which also have restrictive anatomic and functional characteristics (leaflet restriction and lack of available leaflet tissues for better coaptation). The use of the Bioring® offers a great advantage, potentially preserving growing of mitral annulus. These encouraging results should be a motivating drive for surgical teams to proceed to early repair of congenital mitral defects in order to preserve left ventricular function.

P-64
The use of biodegradable rings in atrio-ventricular valve reconstruction
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Introduction or Basis or Objectives: Biodegradable annuloplasty rings promote creation of scar that reinforces valve annulus and enables its growth capacity. Preliminary results of biodegradable rings application during reconstruction of atrio-ventricular valves in children are reported.
Methods: In period from 2005-06-01 to 2006-12-21 18 children were operated for severe insufficiency of A-V valves: Ebstein's anomaly (6pts.), TI (7pts.: 4pts. – redo ToF, 1pt. – redo ASD I; 1 pt. – redo VSD and 1p. – TV dysplasia), MI (5pts.: 3pts. – redo ASD; 1pt. – HOCM+MI; 1p. – ASD I). Median age was 13.6 years (1.3–18.5 years). Median weight and BSA were respectively: 44 kg (8.6–105 kg) and 1.42 m^2 (0.42–2.4 m^2). Type of procedure depended on specific CHD including valve reconstruction with the use of biodegradable ring (Bio-Ring). The effect of operation was evaluated intraoperatively by means of TEE. All patients were followed-up by TTE at discharge and 1.6 and 12 months after the procedure.
Results: No early and late deaths were observed. Median ECC time was 93min (37–230min), median aortic cross-clamp time was 49min (0–120min – 2 pts. operated on beating heart). Implanted ring diameters were from 18 to 34mm. Median follow-up time was 190 days (7–367days). Median tricuspid valve insufficiency fraction dropped significantly ($p < 0.05$) from 39.2% (15.6–93%) to 11.1% (0–28.8%) at discharge and was respectively 14% (3–40.6%), 16.5% (7.5–29%) and 11.7% (5.8–22%) at 1.6 and 12 months after the operation. Median mitral valve insufficiency fraction decreased significantly ($p < 0.05$) from 67% (37.2–80%) to 9.5% (4.9–50%) at discharge and was respectively 8% (4.9–15%) and 13.7% (10.4–17%) at 1 and 6 months after the operation. No median pressure gradient across corrected valves exceeded 10 mmHg throughout entire follow-up. Changes in median BSA and postoperative median valve diameter as well as in resulting valve diameter to BSA index were not statistically significant.
Conclusions: The implantation of biodegradable rings is safe. Enables easy reinforcement of the native valve ring. Ring implantation decrease operation time comparing to classical devices. Early and short-term results are good in the context of corrected congenital heart defect spectrum. Further observation of larger cohort of patients is required in order to evaluate valve annulus growth capacity.

P-65
N-Terminal Pro-Brain Natriuretic Peptide as a Perioperative Biomarker in Paediatric Cardiac Surgery: Preliminary Results
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Introduction: N-terminal pro-brain natriuretic peptide (NT-proBNP) is well correlated with morbidity and mortality in adult cardiac patients. Many studies regarding this subject have been published but only a few were related to perioperative course, especially in children.
Objectives: To assess the association between NT-proBNP plasma levels and outcomes or events after cardiac surgery in paediatric patients.
Methods: Prospective, observational study including 37 children undergoing cardiac surgery. Preoperative, intraoperative and postoperative data were collected, including NYHA classification, echocardiografic study, duration of cardiopulmonary bypass and aortic cross-clamp time. NT-proBNP plasma levels were measured before surgery, 12 hours after surgery and before discharge from the intensive care unit (ICU). Outcomes were assessed by duration of mechanical ventilation, inotropic therapy and ICU stay and the presence of major cardiac events.
Results: From 37 children initially enrolled in the study, three newborns were excluded because of the great variability of NT-proBNP in this age group. The 34 children included were classified in two groups, group I – without major cardiac events (n = 29; mean age: 6.79+/−5.12 years; mean weight: 23.0513+/−.99 kg; mean postoperative NT-proBNP: 3851.295+/−541.45 pg/ml) and group II – with major cardiac events (n = 5; mean age: 2.684.1+/− 4 years; mean weight: 12.761+/−1.77 kg; mean postoperative NT-proBNP: 26533.814+/−023.58 pg/ml). There was no statistically significant difference in age and weight between the two groups ($p = 0.100$ and 0.102 respectively).
Postoperative NT-proBNP plasma levels in group II were positively associated with: mechanical ventilation >12 h ($p = 0.000$), duration of inotropic therapy >48 h ($p = 0.000$), duration of intensive unit care stay > two days ($p = 0.000$). There was no relation between postoperative NT-proBNP plasma levels and duration of cardiopulmonary bypass ($p = 0.722$) and aortic cross-clamp time ($p = 0.880$).
Conclusions: In this study elevated postoperative NT-proBNP plasma levels were associated with major cardiac events. NT-proBNP may be a useful marker in risk stratification of children undergoing elective cardiac surgery.

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P-66

Hemodynamic and oxygenation changes after oral Sildenafil for pulmonary hypertension in infants and children

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Objectives: The immediate effects on hemodynamics and oxygenation after oral Sildenafil are poorly known, and were studied in our tertiary referral centre.
Methods: Between 2003 and 2006, 32 patients with pulmonary hypertension received 35 courses of oral Sildenafil (starting dose 0.25 mg/kg/dose to a maximum of 2 mg/kg/dose QED). Heart rate (HR), systolic, mean and diastolic blood pressures (SBP, MAP, DBP), central venous pressure (CVP), pulmonary artery pressure (PAP) (when invasive monitoring was obtainable) and transcutaneous oxygenation saturation (tcSat) were retrospectively studied. Timepoints were baseline (immediately before), 1, 6, 12, 24 and 48 hours, respectively after the first dose of sildenafil. Median age was 149 days (range 7 days–16.5 years). Patients were allocated to different groups: medical (MED; 6 courses), congenital heart disease (CHD; Sildenafil treatment not concomitant to surgery; 8 courses), and surgical (SURG; first Sildenafil treatment immediately after surgery on cardio pulmonary bypass; 21 courses). Patients were also grouped according to ventilatory support: inhaled nitric oxide (iNO; 15 courses) conventional ventilation without iNO (CV; 7 courses) and spontaneous breathing (SB; 13 courses). Analysis of variance was performed for repeated measurements (significance level $p < 0.05$).
Results: HR decreased in the SURG-group from 142/min to 133/min (p 0.0037) and in the iNO-group from 142/min to 134/min (p 0.0399). SBP decreased from 78 mmHg to 69 mmHg in the iNO-group (p 0.0092). MAP decreased in the SB-group from 69 mmHg to 57 mmHg (p 0.0233). In all other groups a decrease for SBP, MAP and DBP was observed without statistical significance. CVP showed no change in any group. Although invasive PAP-monitoring was only obtainable in 5 patients, it showed a decrease from 34 mmHg to 27 mmHg (p 0.0064). TcSat decreased in the SB-group from 89% to 85% (p 0.049).
Conclusions: Oral Sildenafil can lead to a decrease in heart rate, blood pressure, pulmonary artery pressure and oxygenation. The decrease in MAP and tcSat in spontaneously breathing patients mandates caution when therapy is initiated on a regular ward. Upon therapy initiation, the hemodynamic and oxygenation side effects should be monitored and compensated for in an intensive care unit.

P-67

Do threshold trend fluctuations of epicardial leads predict pacing and sensing characteristics?

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Introduction: AutoCapture devices store diagrams of long-term ventricular pacing threshold trends.
Aim of this study was to evaluate whether the magnitude of ventricular threshold fluctuations (Δfluctuation) predict ventricular threshold and sensing changes over time.
Patients and Methods: A total of 56 children (age 6.7 years (0.0–17.7) received AutoCapture devices connected to bipolar steroid-eluting epicardial leads (Medtronic CapSure Epi 10366 or 4968). For the suture of the leads, a single knot technique was used. Date of lead implant was set as day 0. Maximum lead age at study closure was 12.2 years (median 4.0). Telemetry data and Δfluctuation of threshold trend graphs with 24-hour sample intervals were obtained every 6 months. Regression slope coefficients and individual mean values of repeated measurements were calculated for each patient's course. Data are given as median.
Results: High Δfluctuation correlated with higher thresholds ($rho = 0.68$, $p < 0.001$), lower impedances ($rho = -0.38$, $p = 0.004$), and a Δfluctuation incline ($rho = 0.34$, $p = 0.01$) over time. Furthermore, a Δfluctuation incline correlated with a pacing threshold incline ($rho = -0.34$, $p = 0.01$). ΔFluctuation did not correlate with ventricular sensing ($rho = -0.13$, $p = 0.36$) or cardiac evoked response signals ($rho = -0.005$, $p = 0.97$). No difference for Δfluctuation was found for the pacing site ($p = 0.75$). ΔFluctuation was higher in children with than without congenital heart disease (0.73 versus 0.41 Volt@0.5ms, $p = 0.015$). A higher Δfluctuation was observed if lead age was >5 years compared to ≤5 years (0.75 versus 0.55 Volt@0.5ms, $p = 0.028$).
Conclusions: High amplitudes of ventricular threshold fluctuation predict higher and increasing pacing thresholds, and lower impedances. Theoretically, this results from electrode micro-instability on the epicardial surface with continuous tissue irritation. A fast conducting electrode-tissue interface, such as local edema can be anticipated as a possible etiology for the correlation between high Δfluctuation and lower impedances. Higher Δfluctuation beyond a lead age of 5 years could represent diminished steroid elution, and therefore reduced protection of the electrode-tissue interface.
Potential implications of marked Δfluctuation are short-term follow up as well as lead replacement at the time of elective device exchange.

P-68

Impantable Cardioverter Defibrillators Safety and Efficacy in Children and Young Adults with Congenital Heart Disease

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Introduction: Implantable cardioverters defibrillators (ICDs) are a life-saving therapy with documented efficacy in adults, mainly with coronary artery disease. However, there is limited experience with ICDs in children and young adults with congenital heart disease (CHD). The purpse of this study was to define the efficacy and safety of ICDs in children and young adults with CHD in our center.
Methods: Review of medical records of pediatric patients or young adults with CHD who underwent ICD implantation from 10/1997–12/2006.
Results: Sixteen patients (4 females), 5–34 (20 ± 8.6) years of age, received an ICD. Implantation was endocardial in 14 and epicardial in 2 pts. Ten pts received a dual chamber and 6 pts a single chamber device. Underlying diagnoses were dilated cardiomyopathy (3), hypertrophic cardiomyopathy (2), long QT syndrome (2), catecholaminergic polymorphic ventricular tachycardia (VT) (3), arrhythmogenic right ventricular cardiomyopathy (2), tetralogy of Fallot (2), Ebstein's anomaly (2). Previous operations included repair of tetralogy of Fallot (2), Fontan operation (1), tricuspid valve replacement (2) and pacemaker implantation (2). Indications

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for implantation were: cardiac arrest (3), syncope (4), sustained VT (5), malignant family history (2), high-risk hypertrophic cardiomyopathy (2). Defibrillation threshold was <10–15 J in all pts. Acute complications included prolonged ventricular fibrillation (VF) (1) and pneumothorax (1). One pt died 24 hrs after implantation from VF storm. During follow-up for 1 month–8 years, 5 pts received appropriate therapies for VT/VF and 3 pts received inappropriate therapies (2 T wave oversensing, 1 atrial tachycardia). There were no other chronic complications and all pts except for the one early death, are alive and stable.
Conclusions: ICDs can be used successfully for the treatment of malignant ventricular arrhythmias in children and young adults with CHD with acceptable safety and efficacy.

P-69
Anti-inflammatory Effects of HMG-CoA Reductase Inhibitors (Statins) on Acute Coronary Arteritis in a Rabbit Model of Kawasaki disease
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Introduction: Recently, it has been demonstrated that HMG-CoA reductase inhibitors (statins) significantly reduce major coronary events. In addition, recent observations also suggest that some of the clinical benefits associated with statin therapy may be pleiotropic; that is, they are independent of their cholesterol-inhibiting action. In this study, we tried to evaluate the availability of statins mediated through their anti-inflammatory effects on coronary arteritis in a rabbit model of Kawasaki disease (KD).
Methods: Japanese white male rabbits (age; 5~7 weeks of age, weight; 900–1,000g) were used in this study. Coronary arteritis was induced by the intravenous administration of horse serum twice into juvenile rabbits. Furthermore, the rabbit models with coronary arteritis were divided into 3 groups as follows: the no treatment group (A), those treated with Fluvastatin (20mg/kg) (B) and those treated with Pravastatin (10mg/kg) (C) daily from the day after the second administration of horse serum. Then the animals were sacrified and the hearts were removed at day 3, 5 and 7 after the second administration of horse serum. To evaluate the anti-inflammatory effect of the statins on coronary arteritis, we investigated the serial process of the histopathological features during the acute phase of the coronary arteritis in these three groups.
Results: In group A, histological examinations demonstrated severe panvasculitis with endothelial destruction, marked mononuclear cell infiltration in all layers and edematous thickening of the medial layer. These inflammatory findings were the most prominent at the day 3 and same to the histopathological features in KD. On the other hand, in both group B and C, the inflammatory findings were significantly suppressed even at day 3 in comparison with those in group A. Edematous thickening in the medial layer was not significant in the groups B and C, too.
Conclusions: In this study, it was revealed that statins had a significant anti-inflammatory effects on acute coronary arteritis shown in KD. Statins have good possibilities of being effective for preventing the development of coronary aneurysmal change.

P-70
Role of single domains of the Staphylococcal adhesin fibronectin-binding protein A: involvement in invasion and activation of human vascular endothelial cells in a model of S. aureus experimental endocarditis
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Staphylococcus aureus is among the most important bacterial pathogens responsible for endocarditis in children and adults. Essential for the infection process is the high propensity of S. aureus to colonize endovascular tissues, allowing these pathogens to spread via the bloodstream to other tissues. Our recent studies emphasize the significant contribution of fibronectin binding protein A (FnBPA) of S. aureus to endothelial cell adhesion in the pathogenesis of endovascular S. aureus infections and demonstrate their role in inducing a variety of proinflammatory endothelial responses resulting in leukocyte accumulation, cell damage and fibrin deposition. The structure of the FnBPA molecule as well as the regions for fibrinogen and fibronectin binding are known.
In the present study we investigated the role of the single domains of the FnBPA molecule concerning endothelial infection and subsequent proinflammatory activation. It should be investigated if the fibronectin binding regions are also responsible for endothelial adhesion and activation.
Single regions of the staphylococcal FnBPA were constitutively expressed in the non-invasive organism Lactococcus lactis by means of gene transfer. Bacterial adhesion of these mutants was studied as well as endothelial activation in terms of ICAM-1 and VCAM-1 expression.
Incubation of endothelial cells with strains containing the parts of the FnBPA molecule which mediate fibronectin binding led to a high endothelial adhesion rate and furthermore to activation of the endothelial cells. The most important region is the CD domain of the FnBPA molecule. If the CD region is not present the A region with its fibronectin binding region mediated similar endothelial adhesion and activation. Further experiments with the mutant strains which miss the fibronectin binding region did not show any relevant endothelial adhesion and activation.
We conclude that the regions of the FnBPA molecule binding fibronectin also mediate adhesion to endothelial cells and their activation. This gives further insights in the bacterial-endothelial interaction concerning the development of potential, alternative, antibacterial treatment strategies to inhibit inflammation, tissue damage and fibrin deposition at the infected endovascular sites.

P-71
Persistent of high body mass index in pre-pubertal obese children is an early determinant of Pulse Wave Velocity elevation, a marker of arterial stiffness
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Background: In severe obese and overweight post pubertal children stiffness of carotid artery has been shown using vascular echography.
Aim: We sought to measure segmental arterial stiffness using tonometry of applanation in pre-pubertal obese children.
Method: Measure of indirect arterial stiffness was performed from contour analysis of arterial waveforms recorded by applanation tonometry using a SphygmoCor device to define pulse wave velocity (PWV) and augmentation index (AIx), both indicators of arterial stiffness in 42 pre-pubertal obese children (age = 8.8 ± 1.5 years, BMI = 25.3 ± 4.6 kg/m^2) and in 20 age, height matched control subjects (8.2 ± 1.6 years, BMI = 20.5 ± 1.6 kg/m^2). Consecutively carotid and femoral blood pressure waveform were recorded. Wave

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transit time between the two sites was calculated by the system software, using the R-wave of a simultaneously recorded ECG as a reference frame. Surface distance between the two recording sites was then measured, to determine PWV. AIx was calculated as augmentation (AG) divided by pulse pressure (PP) × 100, with AG defined as the contribution that wave reflection makes to systolic arterial pressure.
Results: In pre-pubertal obese children PWV was higher but not statistically significant compared with control subjects (respectively $4.35 \pm 0.60\,m.s^{-1}$ vs $4.24 \pm 0.65\,m.s^{-1}$). AIx was not different between obese children compared with lean subjects (respectively 1.20 ± 13.40 vs 1.30 ± 16.0). PWV was correlated positively and independently with BMI in obese group ($r = 0.56$, $P = 0.006$) but not with age and height.
Conclusion: Pre-pubertal obese children have neither a discernible arterial stiffness nor an increased wave reflection. However, the persistent increase of BMI appears to be a main determinant of PWV which has to be considered as an early powerful independent predictor of arterial stiffness.

P-72
Expression of two-pore domain channel genes in TASK-1 deficient and wild type mice quantified by real time polymerase chain reaction
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Introduction: Potassium channels constitute the largest family of ion channels and are essential for many cellular processes. Two pore domain potassium channels (K2P channels), which are the most recently discovered potassium channels, are characterized by their activity at all membrane potentials. TASK-1, which is predominantly expressed in heart and brain, is inhibited by hypoxia, platelet activating factor and volatile anesthetics. Inhibition of TASK-1 in single cells prolongs the action potential duration and results in early afterdepolarization, which may provoke significant arrhythmias in vivo. In the course of cardiac phenotyping the TASK-1 deficient mouse, we studied the distribution and expression level of K2P channels in the heart of TASK-1 deficient and wild-type mice.
Methods: Following total RNA extraction from left and right ventricles of TASK-1 deficient and wild-type mice ($n = 4$/group), the level of gene expression was measured by real-time SYBR® RT-PCR reagent. Nine K2P channels (TASK-1, TASK-2, TASK-3, TASK-5, TWIK-1, TWIK-2, THIK, TREK-1, TRAAK) were quantified using the mouse ß-glucuronidase (GUS) housekeeping gene as reference. The specificity of the amplified products was examined by dissociation curve analysis.
Results: In the left and right ventricles of wild-type mice TASK-1, TWIK-2 and TREK-1 are predominantly expressed (112%, 43% and 16% expression of GUS). In the TASK-1 deficient mice, there was no significant difference in TWIK-2 and TREK-1 expression compared to the wild-type littermates. The expression of TASK-1 was not detectable. There was a low expression of TWIK-1, THIK, TASK-2, TRAAK and TASK-3 (5%, 4%, 3%, 3% and 1% expression of GUS) in both ventricles of wild-type and TASK-1 deficient mice. TASK-5 expression was not significantly detectable in the ventricles of the wild-type hearts (0.35% of GUS).
Conclusions: Besides TASK-1 other members of the K2P channel family are significantly expressed in the ventricles of mice, with TWIK-2 and TREK-1 being most predominant. There was no significantly different expression pattern in the ventricles of wild-type mice when compared with the expression pattern in the TASK-1 deficient mice. Therefore, TASK-1 deficiency does not result in a compensatory upregulation of the K2P channel genes tested. *(Supported by a grant from the University Duesseldorf)*

P-73
Moderate hypothermia suppresses inflammatory response and cellular activities in human endothelial cells
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Objectives: Hemodyanmic alterations and endothelial dysfunction may contribute to different morbidities after cardiac surgery in infants and children. Hypothermia is still the standard method for organ protection during cardiac surgery by means of cardiopulmonary bypass (CPB). However, the morphological and biochemical mechanisms underplaying the protective mechanisms by hypothermia have not been clearly elucidated. We evaluated different hypothermia-induced extra cellular signals which are transduced to the nucleus by mitogen-activated protein kinases (MAPKs). Therefore we investigated the role of MAPKs, MEK and extracellular signal-regulated kinase (ERK) and in addition intracellular NF-kappaB activation and IL-6 expression under hypothermic conditions.
Methods: Primary cells were isolated from normal human umbilical cord veins (HUVEC) and kept under moderate hypothermia (32°C) and 37°C over a time curse of 48 h. Cell viability and proliferation (trypan blue exclusion and BrdU-Assay), morphological analyse (CD 31 immuncytochemical staining) and cell activation: IL-6 expression and ERK, MEK and Ikappa-Balpha phosphorylation (ELISA and Western blot technique) were investigated. Furthermore HUVEC were stimulated with 500 U/ml TNF-alpha to mimic inflammatory response after cardiopulmonary bypass.
Results: HUVEC showed significantly less cell proliferation under 32°C after 48 h compared to 37°C in TNF-alpha stimulated and non-stimulated cells. Moreover moderate hypothermia (32°C) caused a down regulation of phosphorylated MEK, ERK and NF-kappaB activation and a significantly reduced IL-6 release in activated HUVEC compared to activated control cells under 37°C. Interestingly there was no difference in cell viability after TNF-alpha–stimulation after 48 h under 32°C compared to 37°C control cells.
Conclusion: Moderate hypothermia (32°C) reduces activation and inflammation in cultured HUVEC via ERK and NF-kappaB signal transduction pathway. Moderate hypothermia after cardiopulmonary bypass operations could lead to less inflammation and may probably reduce the risk of postoperative endothelial cell dysfunction.

P-74
Cytokines, matrix metalloproteinases and tissue inhibitors of metaloproteinases in children after cardiac surgery
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Introduction: The inflammatory responses to cardiac surgery have not been thoroughly investigated. Matrix metalloproteinase-9 (MMP-9) and metalloproteinase-2 (MMP-2) are enzymes involved in cytokine processing and leukocyte extravasation. To explore metalloproteinases activation during cardiac surgery we investigated MMP-9, MMP-2, TIMP-1 and TIMP-2 levels in young children during and after surgery. Additionally, we evaluated the perioperative time course of cytokines.
Methods: The investigations were carried out in 23 children, aged 6–23 months undergoing cardiac operation with CPB (CPB group) and 28 children, aged 8–45 months undergoing cardiac operation without CPB (CS group). Studied markers were measured before induction of anaesthesia, at the initiation of CS/CPB, after 30 minutes of CPB, at the end of CS/CPB, and 4 hours, and 48 hours after CS/CPB. Cytokines: IL-8, -1a, -6, -10, -12p70, TNF-b, and MMP-s were determined by flow cytometry and ELISA method.
Results: MMP-9 concentration increased at the end of CPB and remained elevated throughout the 24 hours in both groups of patients. In CPB group the concentration of MMP-9 detected at the end of CPB positively correlated with time of CPB ($r = 0.66$, $p = 0.0004$). TIMPs concentrations decreased significantly after 30 minutes of CPB, remained lowered to the end of CPB, and returned to the start of CPB level at 48 hours. In both groups of patients the serum levels of IL-6 increased dramatically after surgery. In CPB group we found an immediate increase in IL-10 level after operation. On the contrary, in CS group IL-10 concentration rose, with highest value 4 hours after the end of cardiac surgery and returned to basic level 48 hours later.
Conclusion: Our data confirm that metalloproteinases and cytokines play an important role in inflammatory complications after cardiac surgery in children. These findings suggest that kinetics of MMPs concentrations in serum after cardiac surgery appear to depend on many factors. We demonstrated the link between CPB duration and the MMP-9 concentration. Future studies will determine whether inhibition of MMPs activity diminishes morbidity in children after cardiac surgery.
The study was supported by the Servier grant and Medical University grant no. 502-15-498.

P-75
Stent implantation in Aortic Coarctation patients is not related with an increased risk of exercise induced hypertension

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Introduction: Stent implantation for treatment of Aortic Coarctation (AoCo) is increasingly used. Traditionally patients (pts) with corrected AoCo remain at risk of developing exercise induced hypertension and it has not been studied if the interposition of a metal prosthesis in the aorta can increase that risk.
Aim: To evaluate whether stent implantation is related with exercise induced hypertension, when compared with other AoCo treatment methods.
Methods: Sixteen pts (mean age 26 ± 7years, 81% male) successfully treated with stent implantation were studied. Ten pts (mean age 34+/−12 years, 70% male) treated surgically or by balloon angioplasty were also studied as a control group. All pts were submitted to a treadmill exercise test to evaluate arterial pressure response to exercise. The AoCo gradient was assessed at rest and immediately after exercise (at 1, 3 and 5 minutes), using the arm-leg blood pressure gradient (Arm-leg Δp) and the Doppler corrected gradient (Doppler Δp) determined on the continuous wave Doppler flow signal. Differences between the two groups were evaluated.
Results: Both groups have an hypertensive response to exercise (see table).

Differences between the two groups

	Stent (n = 16)	Control (n = 10)	p
Rest Systolic Arterial Pressure	155 ± 27	153 ± 25	0.814
Peak-exercise Systolic Arterial Pressure	216 ± 28	212 ± 29	0.710
Rest Arm-leg Δp	0 (0–14.5)⋆	0 (0–9)⋆	0.737
1 minute pos-exercise Arm-leg Δp	37.9 ± 22.0	39.7 ± 23.0	0.849
Rest Doppler Δp	17.3 ± 10.2	18.3 ± 8.0	0.802
1 minute pos-exercise Doppler Δp	34.2 ± 21.3	39.9 ± 20.1	0.575

All values in mmHg; ⋆ Median and 25th–75th percentile

Conclusions: Implantation of an aortic stent to treat AoCo does not seem to constitute an additional risk factor for exercise induced hypertension when compared with other forms of treatment. There were no significant differences between the two groups at rest or during exercise.

P-76
Inhaled Iloprost Therapy in Children with Pulmonary Arterial Hypertension

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Introduction: Inhaled iloprost (iILO) is approved for adults with pulmonary arterial hypertension (PAH); however, data in children are lacking.
Methods: Twenty three children (median age 11.7 years; range, 5 to 18 years) with PAH (idiopathic, n = 12; congenital heart disease, n = 11) were treated with iILO. Concomitant therapies included iv treprostinil (n = 5), iv epoprostenol (n = 3), iv iloprost (1), subcutaneous treprostinil (n = 1) sildenafil (n = 19), bosentan (n = 14), and calcium channel blockers (n = 4). Patients were studied with lung function testing before and after iILO (n = 14). During chronic iloprost therapy, follow-up included 6 minute walk distance (6 MWD), NYHA functional class and pulmonary function tests.
Results: Acute administration of iILO lowered mean pulmonary artery pressure from 61 ± 18 mmHg to 53 ± 22 mmHg ($p < 0.05$). Acute iILO reduced FEV1.0 and FEF25–75% by 4 and 11%, respectively ($p < 0.05$ for each), with 5/14 patients (36%) showed >15% decrease in airway flow, suggesting acute bronchoconstriction. The median duration of iloprost therapy was 0.9 years (range 0.1–7.9 years). At initiation (n = 23) the median dose of iloprost was 5 μg (range 0.63–10 μg) with a frequency of 6 times daily (range 4–9). At 12 months (n = 12), the median dose was 5 μg (range 2.5–10 μg) with a median frequency of 6 times daily. During chronic iILO, 6MWD increased in 36%, decreased in 7%, and remained unchanged in 57%. At 6 months functional class improved in 33%, decreased in 15%, and remained unchanged in

52%. Sixty-five% of patients continued successfully on long-term iloprost therapy, but 35% stopped iloprost due to lower airways reactivity, clinical deterioration, or death. 8/9 patients transitioned successfully from intravenous prostanoids to iILO, which was sustained during follow-up

Conclusions: iILO acutely improved pulmonary hemodynamics and caused sustained functional improvement in some children with PAH. However, iILO can induce acute bronchoconstriction in some children. Around 65% of the patient remained on iloprost and it was generally well-tolerated but clinical deterioration, side effects and poor compliance due to the frequency of treatments may limit chronic treatment in children. Larger clinical trials are necessary.

P-77

Prenatal echocardiographic diagnosis of congenital heart disease: quid prodest?

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Objective: Our work aims to present the most recent experience of our single center in the prenatal diagnosis of CHD, and to discuss its impact on the postnatal outcome of the affected subjects, in comparison to the outcome of neonates with CHD referred to our center in the same period, without a prenatal diagnosis.

Methods: we reviewed the clinical records of 282 fetuses with a prenatal diagnosis of CHD, from 2631 fetal echocardiograms performed between January 2002 and December 2005. We compared their postnatal outcome with the outcome of 270 neonates with major CHD (group B) among 1138 neonates without a prenatal diagnosis, admitted in the same period for cardiac malformations, excluding those with VSD or other mild CHD not detectable in uterus.

Results: among the 282 fetuses, termination of pregnancy was performed in 56 cases (19.9%), fetal demise occurred in 16 cases (5.7%), 5 were lost at the FU. Among the 205 neonates with prenatal diagnosis (group A), an extra cardiac anomaly or a genetic or chromosomal aberration were present in 34% of the cases, and in 25% in group B (ns). Pretreatment death occurred in 8/205 (3.9%) in group A and in 8/270 (3%) in group B. Neonatal surgery was performed in 108 pts in group A, on pump in 71, off pump in 37; and in 100 in group B, on pump in 58, off pump 42. Surgical complications occurred in 69 pts (63%) in group A and in 59 (59%) in group B. Early surgical mortality was 19% (21) in group A and 13% (13) in group B (ns). Twenty-six patients in group A and 32 in group B had a diagnostic (12 vs. 9) or interventional (14 vs. 23) cardiac catheterization, without mortality or major morbidity.

In conclusion, comparing 2 groups of neonates with major CHD, treated in a short period of time in a single center, in a region where there is a high level of neonatal care, we could not detect any statistically significant difference in the postnatal outcome between the group with and without prenatal diagnosis.

P-78

Tissue Doppler Imaging indices of systolic and diastolic function in the normal fetal heart

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Objective: To establish gestational age-specific values of systolic and diastolic longitudinal wall-motion velocities by spectral Doppler Tissue Imaging (DTI) in healthy singleton fetuses.

Methods: Ventricular myocardial velocites (peak myocardial velocities during early diastole – Em, atrial contraction – Am and peak systole in ejection phase – Sm) were assessed by DTI (17–34 week's gestation) at the right ventricular wall (RV) in 62 fetuses, at the left ventricular wall (LV) in 32 fetuses and interventricular septum (IVS) in 28 subjects. Examinations were performed using Sequoia 512, with cardiological probe of 2.5–7 MHz and pediatric cardiology software. The sample volumes were placed in the basal part of RV, LV wall and IVS in the 4-chamber view.

Results: There was a strongly positive correlation between Em, Am and Sm of RV, LV and IVS and gestational age ($p = 0.001$), Table. Em, Am and Sm were higher at the RV comparing to LV and IVS, Table.

weeks	17–20			21–24			25–29			30–34		
DTI (cm/s)	Em	Am	Sm	Em	Am	Sm	Em	Am	Sm	Em	Am	Sm
RV	4.2	6.9	4.4	4.7	7.9	4.9	5.8	8.9	5.5	6.9	9.4	6.3
LV	3.5	6.0	3.3	3.6	6.1	3.8	4.5	7.5	5.4	5.4	7.6	6.2
IVS	3.3	4.5	2.9	3.4	4.6	3.0	4.0	5.1	3.3	4.1	5.2	4.3

Conclusions: Systolic and diastolic myocardial lengthening improves with advancing gestation. Fetal diastolic function is predominantly contributed by atrial contractions. The highest wall-motion velocities are recorded from RV wall. Assessment of fetal systolic and diastolic function by DTI is technically difficult, especially for LV wall and IVS. DTI requires good echocardiographic skills of performing sonographer, but it may represent a new modality for evaluation of fetal well-being.

P-79

Bruce Treadmill Test in Children: First Normal Values for Children from Western Europe

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Introduction: One of the main goals for exercise testing in children is evaluation of exercise capacity. There are many testing protocols, but the Bruce treadmill protocol is widely used among pediatric cardiology centers. Thirty years ago, Cuming et al. were the first to establish normal values for children from North America (Canada) aged 4 to 18 years old. No data was ever published for children from Western Europe. Our study aimed to assess the validity of the normal values from Cuming et al. for children from Western Europe in the 21st century.

Methods: It is a retrospective cohort study in a tertiary care children's hospital. 144 children referred to our institution but finally diagnosed as having a normal heart underwent exercise stress testing using the Bruce protocol between 1999 and 2006. Data from 59 girls and 85 boys aged 6 to 18 were reviewed. Mean endurance time (ET) for each age category and gender was compared with the mean normal values from Cumming et al. by an unpaired t-test (Fig.).

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Gender	Age Group (yr)	Study	Mean ET (SD)	p
Girls	6 to 7	NA*	11.2 (1.5)	NS
		WE*	11.3 (0.9)	
	8 to 9	NA	11.8 (1.6)	NS
		WE	13.0 (2.3)	
	10 to 12	NA	12.3 (1.4)	NS
		WE	12.6 (2.5)	
	13 to 15	NA	11.1 (1.3)	NS
		WE	12.8 (2.0)	
	16 to 18	NA	10.7 (1.4)	NS
		WE	13.4 (4.7)	

Gender	Age Group (yr)	Study	Mean ET (SD)	p
Boys	6 to 7	NA	11.8 (1.6)	NS
		WE	10.9 (1.3)	
	8 to 9	NA	12.6 (2.3)	NS
		WE	12.4 (1.2)	
	10 to 12	NA	12.7 (1.9)	p < 0.05
		WE	14.0 (2.4)	
	13 to 15	NA	14.1 (1.7)	p < 0.05
		WE	15.3 (2.0)	
	16 to 18	NA	13.5 (1.4)	p < 0.01
		WE	15.5 (1.1)	

* NA : study from Cuming et al for children from North America
* WE : our study for children from Western Europe
NS : non significant – SD : standard deviation

Results: Mean ET increases with age until 15 years old in girls and then decreases. Mean endurance time increases continuously from 6 to 18 years old in boys. The increase is more pronounced in boys than girls. In our study, a significant higher mean ET was found for boys in age categories 10 to 12, 13 to 15 and 16 to 18. No significant difference was found in any other groups.
Conclusions: Some normal values from Cuming et al. established in 1978 for ET with the Bruce protocol are probably not appropriate any more today for children from Western Europe. In 2001, Ahmad et al. showed that non-obese american children and adolescents had lower ET than previously reported by Cuming et al. On the contrary, our study showed that mean ET is not lower, but rather higher for boys from 10 to 18 years old. Despite common beliefs, cardiovascular conditioning doesn't seem yet reduced in children from Western Europe. New data for Bruce treadmill exercise testing for healthy children, 4 to 18 years old, living in Western Europe are required.

P-80
Serum cardiac Troponin T levels in active carditis due to acute rheumatic fever

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Introduction: Acute rheumatic fever (ARF) remains as an important health problem for many developing countries. Carditis is the major form leading to morbidity and mortality. In this study it is aimed to determine the role of cardiac Troponin T (cTnT) levels in patients with active carditis.
Methods: Twenty eight patients (19 male and 9 female) with active carditis according to Jones criteria and 16 healthy children as the control group were enrolled in the study. The mean age for the patient group was 11.1 ± 2.4 years. Serum cardiac cTnT and CK-MB levels were detected by Cardiac T Quantitative device and "cardiac reader" by Boehringer Mannheim and ELISA respectively in the patient group before the treatment and in the control group.
Results: Of the 28 patients with ARF 13(47%) had severe carditis. Serum cTnT levels were within normal limits except one patient with severe carditis who had mitral and aortic regurgitation as well as mild pericarditis (0.26 ng/ml). Serum cTnT levels showed an statistically unsignificant increase in the patients with severe carditis when compared to mild carditis. CK-MB levels were all within normal limits in all patients.
Conclusion: Serum cTnT level is accepted as a sensitive biomarker of cardiomyocyte damage. Though nearly half of the patients had congestive heart failure, normal values of cTnT showed that even intense inflammation in ARF did not lead to myocardial damage.

P-81
The influence of cardiosurgery procedures on the lymphocyte subpopulations in children with congenital heart defects operated in first year of life

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Children with congenital heart defects have increased risk of developing severe infections due to possible impairment of the immune system. Patients undergoing open heart surgery with cardiopulmonary bypass (CPB) usually have partial or total thymectomy to facilitate cannulation of great vessels.
The objective of our study was to investigate if cardiac surgery with CPB has the impact on the circulating lymphocyte subpopulations. Twenty children in first year of life (7 newborns and 13 infants) with congenital heart defect were enrolled and prospectively studied. The percentage of T-cell subtypes (CD3, CD4, CD8), B-cells, NK-cells was determined. In addition in some patients further testing of CD4 and CD8 cells was performed (CD45RA+, CD45RA+) to determine whether they were naive or educated memory lymphocytes. Peripheral blood samples were drawn before surgery, in early postoperative period (mean 8.7 days) and later after surgery (mean 8 months).
The following results were obtained: the mean percentage of CD3 cells early after cardiosurgery procedure was lower than before (59.9% ± 11.4% v. 52.95% ± 14.8%) and dropped significantly in observation period (52.95% ± 14.8% v. 44.45% ± 14.1%; p = 0.002). The changes in CD4 subset were similar (45,5% ± 12,01% v. 42.55% ± 9.54% v. 27.95% ± 10.88%), significant reduce we observed in long-term after surgery (p = 0.0002). In the same period percentage of CD19 cells increased significantly (23.1% ± 8.83% v. 34.95% ± 12.45%; p = 0.0004). CD4/CD8 ratio and NK-cells percentage changes were not significant. The mean percentage of CD4CD45RO+ and CD8CD45RO+ lymphocytes dropped after cardiosurgery not significantly (8.38% ± 3.18% v. 6.43% ± 2.68% and 3.15% ± 2.23% v. 2.71% ± 2.3%). In next months we observed the significant rise in this subsets (6.43% ± 3.18% v. 12.25% ± 4.93%; p = 0.003 and 2.71% ± 2.3% v. 7.2% ± 6%; p = 0.03). The percentage of CD4CD45RA+ and CD8CD45RA+ cells in first days after operation changed not significantly, but in next months significantly dropped (55.31% ± 17.34% v. 41.7% ± 15.85%; p = 0.03 and 30.61% ± 15.92% v. 23.9 ± 5.97%; p = 0.03).
Our observations suggest that cardiosurgery with CPB in young children causes changes in lymphocyte subpopulations in short and in long postoperative period. It could influence immune system function later in life. Systematic evaluation of immune competence should be performed in children undergoing open heart surgery due to congenital anomalies.
The study was supported by grant of Ministry of Science and Higher Education no. 1198/P01/2006/31.

P-82

Assessment of left ventricular systolic function with Tissue Doppler Imaging and conventional echocardiographic methods in children after the successful repair of aortic coarctation

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The aim of the study was an assessment of left ventricular systolic function with Tissue Doppler Imaging (TDI) including strain and strain rate and with conventional echocardiographic parameters in children after the successful repair of aortic coarctation (CoA).
Study group (CoA Patients) consisted of 32 children with the mean age 12.01 ± 4.24 years after the successful surgical repair of CoA. The mean age at the surgery was 3.84 ± 4.04 years and the mean follow-up 8.17 ± 3.29 years. Control group (Controls) consisted of 34 healthy children. According to standard blood pressure measurement study group was divided in to hypertensive (CoAHT Patients) and normotensive patients (CoANT Patients). In TDI the systolic velocity of mitral annulus motion (S'MAM), maximal global systolic strain (Sε) and systolic strain rate (SSR) were analysed. Sε and SSR were evaluated off-line and calculated as a mean values of local strain and strain rate parameters for medial segments of septal, lateral, inferior and anterior left ventricular walls. Moreover the shortening fraction (SF), ejection fraction (EF), mean circumferential fiber shortening (Vcf) and heart-rate corrected mean circumferential fiber shortening (Vcfc) were evaluated. The results were compared between CoA Patients and Controls and between CoAHT Patients and CoANT Patients with Student's t-test.
Results:

	CoA Patients	Controls	p	CoAHT Patients	CoANT Patients	p
S'MAM (cm/s)	6.92 ± 0.75	6.45 ± 0.83	<0.05	6.68 ± 0.73	6.96 ± 0.76	NS
Sε (%)	−28.67 ± 6.04	−22.53 ± 6.44	<0.001	−28.18 ± 4.80	−29.09 ± 6.47	NS
SSR (s^{-1})	−3.20 ± 0.76	−2.39 ± 0.49	<0.001	−3.19 ± 0.39	−3.20 ± 0.84	NS
SF (%)	41.65 ± 6.26	38.72 ± 4.76	<0.05	42.21 ± 10.07	41.46 ± 4.68	NS
EF (%)	79.48 ± 6.10	76.58 ± 5.46	<0.05	79.22 ± 9.37	79.57 ± 4.83	NS
Vcf (circ/s)	1.45 ± 0.27	1.01 ± 0.26	<0.001	1.31 ± 0.23	1.50 ± 0.28	NS
Vcfc (circ/s)	1.31 ± 0.24	0.93 ± 0.19	<0.001	1.26 ± 0.24	1.33 ± 0.24	NS

Conclusions: 1. In children after the successful surgical repair of aortic coarctation left ventricular systolic performance remains increased in late follow-up in hypertensive as well as normotensive patients. 2. Higher systolic strain and strain rate in children treated due to aortic coarctation may indicate the rise of left ventricular myocardial contractility. 3. Tissue Doppler Imaging is a simple and useful method in assessment of left ventricular systolic function in children after the surgical repair of aortic coarctation.

P-83

Acute heart disease following Chikungunya virus infection in the neonate

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Background: In 2006, Reunion Island, a South West Indian Ocean French department, experienced a massive outbreak of Chikungunya, an arbovirus (alphavirus) infection transmitted by mosquitoes (Aedes Albopictus).
Methods: Between January and July 2006, 19 neonates were admitted for severe Chikungunya virus infection (CHIKV) in our neonatal intensive care unit. Virological confirmation was obtained by specific IgM and/or genome RTPCR detection in blood. All patients were evaluated for cardiac involvement by clinical examination, 12 leads ECG, 2D echo, biochemistry for CPKMB, ASAT, Troponin Ic and ProBNP. Description and outcome are reported.
Results: 8 patients (7 male, 1 female) had cardiac complications. Mean age of onset for symptoms was 8 days (2 to 30). Severe neurological disorders were associated in 6 cases and hemorrhagic symptoms in 2 cases. In 6 patients, mother to child transmission of CHIKV was suspected. Three patients had pericardial effusion, 7 myocarditis (left ventricular cardiomyopathy with elevated cardiac enzymes) and 1 had ventricular arythmias. Coronary artery dilatation with parietal thickening but without aneurysms was observed in 6 cases. Outcome was favorable in all patients. After 6 months, three patients had an hypertrophy of left ventricle and one a moderate dilatation of coronary arteries.
Conclusion: CHIKV can be associated with cardiac complications in the neonate, either isolated or associated with other severe anomalies. Remodeling of the coronary arteries, resembling Kawasaki or herpes virus infections can be observed on 2D echo. Short term evolution was favorable in our experience but long term follow-up is recommended.

P-84

Register of congenital heart defects (CHD) in Croatia (Croatian study – October 2002–January 2007)

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Introduction: Since 1979 congenital heart defects (CHD) have been followed in Europe by EUROCAT study (European Registry Of Congenital Anomalies and Twins). EUROCAT study is population register of CHD based on 21 regional center in 11 european countries and includes 240 000 labors per year. We formed Register of CHD in Croatia on EUROCAT study model because of high quality of their pattern and our goal to be part of EUROCAT study some time in the future. It is formed not only as population register but also as hospital register that includes details of patient diagnoses, treatment and enables us to make life-survival curves.
Methods: We formed the Register including all children with CHD treated in 15 institutions in Croatia, born since December 2002 as a prospective study according to EUROCAT study model.
Results: Since December 2002, 1492 children with congenital heart defect was recognized, female 719 (48%) and 773 (52%) male. According to anamnestic data 4% of them was born from pregnancy complicated with acute infection, 2% from mothers with chronic diseases (diabetes mellitus, epilepsy, mesenhymopathias), 2% have CHD in family and 2% other inherited diseases. Out of 1492 children 432 (29%) had ventricular septal defect, 334 (22%) atrial septal defect, 167 (11.2%) patent aterial duct, 80 (5.4%) pulmonary valve stenosis, 35 (2.3%) aortic valve stenosis, 36 (2.4%) tetralogy of Fallot, 44 (2.9%) transposition of great arteries, 48 (3.2%) coarctation of the aorta, 48 (3.2%) atrioventricular canal, 25 (1.7%) hypoplastic left heart syndrome, 22 (1.5%) cardiomiopathies, 50 (3.4%) arrhythmias and 60 (4.1%) complex heart malformations or deformations. Operation underwent 273 (18.3%), 256 (17.2%)

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was treated only medicamentously. Extracardiac anomalies was recognized in 206 (13.8%) children and among them 73 (35.4%) with Down syndrome. Only 32 (2.1%) were diagnosed prenataly. During follow up 43 (2.9%) children died (17 of them were operated).
Conclusion: The incidence of CHD in Croatia (8 0/00) corelates to world data as well as the most common diagnoses,VSD followed by ASD. The aim of Register is to establish base for follow up of children with CHD for whole country.

P-85 withdrawn

P-86
A rare cause of mitral involvement in the children: the Geleophysic Dysplasia (GD)
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Introduction: In pediatric population most cases of mitral involvement have congenital etiology. In developed countries acquired mitral disease are rare and they have generally rheumatic origin. Mitral dysplasia may be pathognomonic of some syndromes or diseases. Geleophysic Dysplasia (geleos-happy; physis-nature) is a rare congenital disease of uncommon origin characterized of abnormal accumulation of storage material in the extracellular matrix. We report a series of patients with Geleophysic Dysplasia affected by progressive mitral involvement.
Patients and Results: Between 1995 and 2007 we identified 7 patients affected by DG having mitral involvement. Among 4/7 patients, echocardiography was normal before clinical diagnosis; in the remaining 3 patients mitral involvement diagnosis suggested diagnosis of GD. Three patients had a mitral disease, 2 patients had mitral stenosis, 1 patient had mitral insufficiency without stenosis. In all patients mitral leaflets were thickened and sclerotic. In 4 patients papillary-commissural fusion was found. Dysplasia involved valves other than mitral in 3 patients. Three patients underwent mitral valve remplacement before the age of 2 years. Two patients died because of intra-operative and post-operative complications.
Conclusions: GD is a rare cause of cardiac involvement in children which should be considered in the differential diagnosis of progressive mitral dysplasia. Prognosis may be poor. Detection of tracheo-bronchial involvement by extracellular infiltration is recommended before to attempt cardiac surgery.

P-87
Children with Congenital Heart Disease: Effectiveness of a Respiratory Synctial Virus Immuno-Prophylaxis Program
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Introduction: Children with congenital heart disease (CHD) hospitalized with Respiratory Syncytial Virus (RSV) have a mortality and hospitalization rate that is significantly greater than children without CHD. We postulated that children with CHD would benefit from RSV immuno-prophylaxis (RSVIP) with decreased RSV-related hospitalizations. A provincial RSVIP program was initiated in the fall of 2003 and provides ongoing identification and tracking of eligible children, family and caregiver education, and monthly injections of palivizumab during the RSV season.
Purpose: To assess the impact on RSV disease severity, we compared a cohort of children with CHD who were admitted to hospital with RSV (Pre-RSVIP cohort) to children with CHD admitted to hospital with RSV after implementation of a RSVIP program (Post-RSVIP cohort).
Methods: Children with CHD who would have been eligible to receive immuno-prophylaxis were treated for RSV infection at a tertiary care pediatric hospital between November 1, 1998 and June 30, 2003 (Pre-RSVIP cohort, N = 20) and November 1, 2003 and June 30, 2006 (Post-RSVIP cohort, N = 5) were included.
Results: The median length of hospital stay (LOS) was longer for the Pre-RSVIP cohort (19 vs 5 days, $P < 0.01$). Twelve children were admitted to ICU with a LOS of 6.5 days in the Pre-RSVIP cohort compared to 1 child in the Post-RSVIP cohort who spent 10 days in the ICU. The Pre-RSVIP cohort required significantly more total days of oxygen therapy (147 vs 14 days, $P < 0.001$) and mechanical ventilation (124 vs 8 days, $P < 0.001$). Seven patients had surgery delayed or expedited as a result of RSV infection in the Pre-RSVIP cohort compared to no patients in the Post-RSVIP cohort. Feeding difficulties, motor delay, and chromosomal abnormalities were common in both cohorts. There were no RSV-related deaths.
Conclusions: There was a 92% relative reduction in hospitalization from the Pre- to Post-RSVIP cohort. The Post-RSVIP cohort had less disease severity, admissions to the ICU, days requiring oxygen therapy and mechanical ventilation, and had fewer children whose surgery was either delayed or expedited. Further study will determine additional risk factors that contribute to the burden of RSV infection in children with CHD.

P-88
N-terminal-pro-B natriuretic peptide levels in patients after Fontan operation correlate with congestive heart failure
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Introduction: Ventricular function is critical in the long-term management of patients with Fontan physiology. Early and minor deteriorations of single ventricular function are difficult to assess.
Purpose of the study: To generate normal values of N-terminal-pro-B natriuretic peptide (NT-pro BNP) in children after Fontan operation and to test the hypothesis that plasma levels of NT-pro-BNP correlate with clinical severity of congestive heart failure (CHF).
Patients and methods: NT-pro BNP plasma levels of 59 patients (median age: 8.4 years, range: 2.12–25 years) after Fontan operation (median time of follow up: 3.4 years, range 0.5–14.7 years) were measured using an automated enzyme immuno assay (Roche Diagnostics). At the same time all patients had a chest x-ray, an echocardiogram and a clinical examination by a pediatric cardiologist. The degree of heart failure was quantified using the New York university pediatric heart failure index (NYPHFI). 24 patients had single right ventricle morphology (RV), 26 patients had single left ventricle morphology (LV) and 9 patients had single ventricles with undeterminated morphology.
Results: NT-pro BNP levels strongly correlated with the NYPHFI ($p = 0.0001$). In patients with CHF (14/59) the NT-pro BNP levels were significantly higher (median 399, range: 140–5440 pg/

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ml) than in patients without CHF (median 96, range: 11–376 pg/ml). The 95th percentile of NT-pro BNP in patients without CHF was 252 pg/ml. There was no significant difference, but a tendency towards higher NT-pro BNP levels in patients with single RV compared to patients with single LV.
Conclusion: Plasma NT-pro BNP levels can help in the detection and assessment of CHF in patients after Fontan operation. Patients after Fontan procedure with NT-pro BNP levels of more than 252 pg/m should be carefully assessed for signs of heart failure. NT-pro BNP levels could be used for monitoring the effect of various treatments for CHF in children after Fontan procedure.

P-89
Sildenafil in Cystic Fibrosis as a Disease Model for Pulmonary Arterial Hypertension Caused by Impaired Gas Exchange
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Introduction: Idiopathic and congenital heart disease related PAHs are predominantly vascular processes that respond well to therapeutic vasodilators. However, if PAH occurs in lung disease such as cystic fibrosis (CF), there is tangible risk that vasodilative therapy will cause unwanted reduced oxygen saturation by increasing intrapulmonary shunting. We describe our experience with pulmonary vasodilative therapy in a girl with CF and right ventricular (RV) failure.
Case Report: We report on a 17-year-old female with CF and chronic hypoxia who was referred because of PAH with RV-failure in clinical function stage NYHA III-IV. The following data were measured during initial presentation: transcutaneous saturation (TCS) 90%, exercise tolerance 1.03W/kg⋆min (30% of predicted), maximum oxygen uptake (VO_2 max) 14.2 ml/min⋆kg (31% of predicted), 6-minute walking distance (6-MWD) 480 m and echocardiographically measured RVSP 75 mmHg. Cardiac catheterisation confirmed increased RVSP of 65 mmHg with end-diastolic pressure of 10 mmHg, mean pulmonary arterial pressure of 45 mmHg and increased PVR of 12WU⋆m^2, while cardiac output was 3.7 L/min⋆m^2.
Treatment Result: Sildenafil therapy 20 mg TID was started. 6 months after initiation of treatment, our patient showed subjective improved quality of life and a function stage NYHA II. Increased values of TCS (96%), exercise tolerance (1.62W/kg⋆min) (46% of predicted), VO_2 max (16.7 ml/min⋆kg) (43% of predicted) and 6-MWD (555 m) could be reported. Repeated invasive measurements showed slightly decreased hemodynamic pressures with a mean pulmonary arterial pressure of 38 mmHg, RVSP of 60 mmHg with end-diastolic pressure of 7 mmHg, and significantly reduced PVR of 6 WU⋆m^2 with increased cardiac output of 5.7 L/min⋆m^2.
Conclusions: This patient of severe CF with RV-failure due to PAH profited from an oral treatment with sildenafil according to clinical, hemodynamic and exercise related criteria. Interestingly and importantly, the treatment was not associated with an increase in intrapulmonary shunting, thus supporting the notion of an intrapulmonary selective action of sildenafil. Further studies on the effect of specific pulmonary vasodilative therapy seem to be warranted to evaluate the benefit in regards to exercise tolerance and overall quality of life in these patients.

P-90
Early detection of myocardial dysfunction in childhood patients with beta-thalassaemia major
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Objective: To evaluate left and right ventricular myocardial performance using pulsed-tissue Doppler imaging (TDI) and its relation to BNP levels in patients with Beta-Thalassaemia Major (ß-TM).
Method: We enrolled 35 thalassaemic patients (21 male, 14 female, and 14.2 ± 4.1 years) with normal left ventricular systolic function and 20 healthy control subjects (11 male, 9 female, and 12.5 ± 4.2). By TDI, the sample volume was placed at the lateral margin of the mitral annulus (Emw, Amw, Smw), interventricular septal annulus (Eivsw, Aivsw, Sivsw), and the lateral margin of the tricuspid annulus (Etw, Atw, Stw). Myocardial performance indexes (MPI) of left ventricular (LV) lateral wall, interventricular septum (IVS) and right ventricular (RV) lateral wall was calculated with TDI. Plasma BNP levels were measured in all patients. MPIs and other echocardiographic parameters of patients with ß-TM were compared with control group.
Results: All the patients' plasma BNP levels were within normal limits. Echocardiographic parameters of the study group are given in Table 1. There were no differences between mean fractional shortening (FS) of LV, transmitral flow velocities, and E/A ratio of patients and control group. MPI of LV, IVS, and RV of patients were significantly higher than control group (p: 0.01 and p: 0.004, and $p < 0.001$, respectively).

Table 1: Echocardiographic parameters of the study group

Parameters	ß-TM	Controls	p
LV FS (%)	35.8 ± 5.03	37.4 ± 4.40	NS
LV IVRT (ms)	56.8 ± 12.67	56.1 ± 8.56	NS
Mitral E/A	1.64 ± 0.27	1.54 ± 0.11	NS
RV IVRT (ms)	44.4 ± 6.21	41.7 ± 5.13	NS
RV E/A	1.68 ± 0,20	1.55 ± 0.21	NS
LV MPI⋆ (TDI)⋆⋆	0.40 ± 0.06	0.36 ± 0.04	0.01
IVS MPI (TDI)	0.40 ± 0.05	0.36 ± 0.04	0.004
RV MPI (TDI)	0.44 ± 0.06	0.38 ± 0.04	<0.001

⋆ Myocardial performance index, ⋆⋆ Pulsed-tissue Doppler imaging

Conclusion: Our study confirms that MPI obtained by TDI seems to be an early sensitive parameter to detect cardiac dysfunction in ß-TM. To our knowledge, this study is the first evaluation of left and right ventricular systolic and diastolic functions using MPI obtained by tissue Doppler echocardiography in ß-TM patients.

P-91
Preoperative mortality in patients with transposition of the great arteries
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Introduction: After institution of the arterial switch operation as the treatment of choice for transposition of the great arteries survival has improved. However, mortality before surgery still remains significant, although reports about mortality before surgery are extremely scarce. We therefore studied all our patients with TGA from 1990 onwards with respect to presurgical death.
Methods: Retrospective review of all patients with TGA that were amenable to biventricular repair from 1990 onwards. We studied

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the time of diagnosis after birth, use of NO inhalation, performance of Rashkind procedure and survival until surgery.
Results: The diagnosis of TGA was made in 158 patients after an average period of 9 ± 18 days (median 1 day) after birth. Patients with an additional ventricular septal defect (n = 26) were diagnosed significantly later (P < 0.01). Of the 158 patients, 10 died before surgery (6.3%). All 10 had simple TGA without additional abnormalities. These 10 patients were highly symptomatic and a diagnosis was made within the first 24 hours after birth. Of the 10 patients 9 died because of pulmonary artery hypertension and profound hypoxemia. One patient died of irreversible shock after necrotizing enterocolitis. Risk factor analysis showed that symptom onset within 24 hours after birth (P < 0.01) and intact ventricular septum (P < 0.05) were independent risk factors for preoperative mortality. When the periods 1990–1997 and 1998–2006 were compared, no differences in preoperative mortality were found. Of the 82 patients during the 1990–1997 period 6 died before surgery (7.3%), and of the 76 patients during 1998–2006 4 patients died (5.3%; all treated with NO ventilation). None of the 10 patients were treated by mechanical circulatory support. Of the overall group of 158 patients only 3 were diagnosed prenatally, one of whom died before surgery because of irreversible pulmonary hypertension.
Conclusions: Preoperative mortality occurs in 6% of all TGA patients. These patients present with severe cyanosis during the first 24 hours after birth. These highly symptomatic infants have a high mortality despite urgent atrial septostomy and NO ventilation. Measures to improve survival to operation may include prenatal diagnosis and even more rapid diagnosis and treatment once these infants become symptomatic.

P-92

Diagnostic value of noninvasive scintigraphy with 99mTc-Anti-Granulocyte antibody in children with clinically suspected myocarditis – a preliminary report

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Objectives: Clinical diagnosis of myocarditis (myo) is difficult. The aim of study was to investigate whether scintigraphy with 99 mTc-Anti-Granulocyte antibody (99mTcAGA) is useful in children with suspected myo and what is its correlation with EMB.
Methods: From 2005 to 2006, 8 children (6 boys, 2 girls; aged 6.6-17.6 yrs; mean 13 ± 3.8 yrs) presenting with symptoms of myo were evaluated at the time of initial presentation and 6 months after the first study. In all pts heart scintigraphy with 99 mTcAGA was performed. Estimation of antigranulocyte antibodies uptake was performed by calculation of the heart-to-lung ratio (HLR), value above 1.50 was used as a positive result. Control group consisted of 10 children without cardiovascular disease underwent scintigraphy with 99mTcAGA due to suspicion of enterocolitis (HLR 1.06–1.50). EMB with immunohistchemical methods was done in 7 pts (no consent for EMB in 1 pt).
Results: The mean time from onset of symptoms of myo to diagnosis was 2.52 ± 3 mo. All pts presented with fatigue, heart failure (n = 4), ventricular arrhythmias (n = 4), chest pain (n = 3). In 7 pts positive antigranulocyte uptake was observed (mean HLR 2.04 ± 0.54; range 1.66 to 3.2). In 3 pts intense uptake was found (HLR > 2), in 4 pts moderate (HLR<2) and in 1 pt borderline (HLR 1.50). The pts who presented within 2 mo of the onset of symptoms demonstrated higher HLR (mean 2.12. ± 0.66) than those presenting during the later phase (mean HLR 1.74 ± 0.07). EMB results in 6 pts showed evidence of myo, in 1 pt no features of myo. All 3 pts who had an HLR > 2 demonstrated in EMB evidence of active myocarditis. Follow-up scintigraphy was performed in 6 pts (mean HLR 2.01 ± 0.53; range 1.45 to 2.9). Follow-up EMB performed in 5 pts showed evidence of persistent inflammatory process in myocardium.
Conclusion: (1) In 83.3% pts with positive scintigrapy results biopsy-proven myocarditis was observed. (2) Intense myocardial uptake of antigranulocyte antibodies with HLR > 2 strongly suggests active myocarditis. (3) Scintigraphy with 99mTcAGA seems to be a useful diagnostic method in children with suspected myocarditis, but further studies are needed to establish its sensitivity and specificity.

P-93

High Rate of Off-Label use in cardiovascular paediatric pharmacotherapy requires new focus in research

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Purpose: In paediatric patients, off-label use of drugs, i.e. their use outside the terms of approved label, is associated with a higher rate of adverse drug reactions and inefficacy of therapy. We performed a study to identify the drug groups which are associated with the highest relevance of off-label use among paediatric inpatients.
Method: Between January and June 2006, prescriptions of 351 patients were collected, documented and analysed on a ward at the University Children's Hospital in Düsseldorf. Diagnoses of patients were obtained from the discharge reports and were classified in groups according to the International Classification of Diseases (ICD-10). The approved labels of drugs were obtained from the summary of product characteristics (website information of manufacturer). Drug groups were classified according to the Anatomical Therapeutic Chemical Classification System (ATC-Code).
Results: We analysed 1540 prescriptions of 191 different drugs. We identified 193 patients (55%) with at least one prescription of an off-label drug. Of all analysed prescriptions, 388 (25%) were off-label. The percentages of off-label use among the most prescribed drug groups were as follows: 56% of the drugs for the cardiovascular system (ATC-Code C and B01: 100 from total 197 prescriptions), 41% of anti-infective drugs (ATC-Code: J, A07 and S01: 157 from 385), 25% of drugs for the respiratory system (ATC-Code R: 72 from 283), 14% of drugs for the gastro-intestinal tract and metabolism (ATC-Code A: 33 from 232) and 3% of analgesics and antipyretics (ATC-code: N02 and M01; 6 from 236).
Conclusion: Authorization of paediatric drugs is required especially in the domain of cardiology. Since the new EU-Regulation on paediatric medicines will lead to a strong increase in paediatric studies in the near future, this result gives an important impulse, as it shows up targets for paediatric clinical studies.

P-94

Peculiar cardiac involvement in the infant of diabetic mothers

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Objectives: To emphasize peculiar aspects of cardiac involvement in infant of diabetic mother.
Material and method: Subjects – 55 newborns of diabetic mothers, follow up over a period of five years. All patients were assessed in the first week of life and 29 of them were evaluated also at 6–12 month, following a protocol which included: clinical examination, ECG, cardiothoracic radiography, and Doppler echocardiography for congenital and/or acquired pathology. Fetal echocardiography was not done in any of pregnancies.
Results: Clinical: in 24 patients a systolic murmur was found, the other being asymptomatic or presenting signs for other pathology than cardiac. Electrocardiography revealed left ventricular hypertrophy in 12 cases and disturbed ventricular repolarization in 27 cases. Cardiomegaly was found in 12 cases on thoracic radiographies. Echocardiography findings were: nonobstructive hypertrophic cardiomyopathy with septal thickness in 39 cases, arterial pulmonary hypertension in 5 cases, left ventricular diastolic dysfunction with normal systolic function in 58% of cases and congenital cardiac malformations as patent ductus arteriosus (5 cases), ventricular septal defect (3 cases) and foramen ovalae (7 cases). At 6–12 months, 22 cases which were diagnosed at birth with cardiac modifications was normal morphological cardiac aspect (15 cases), or significant reduction of hypertrophic cardiomyopathy (7 cases), all of them showing normal diastolic and systolic left ventricular function.
Conclusions: Infant of diabetic mother presents a high risk for cardiac involvement, either cardiac congenital malformations (27% of cases) or acquired cardiac pathology-hypertrophic cardiomyopathy (71% of cases) which justifies early cardiologic screening for all of these newborns in presence or absence of cardiac suffering signs or symptoms. Echocardiography is the most sensitive method of diagnostic, noninvasive, useful for primary diagnostic as well as for follow up.

P-95
Congenital complete AV block in children – clinical course and outcome
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Objective: To search for peculiarities in the clinical course, indications for pacemaker implantation (PM) and prognosis according to the age at diagnosis in patients with complete congenital AV block (CCAVB).
Patients: Forty-five children with CCAVB were studied with a mean follow-up period of 8.56 ± 6.3y (0.2–21.6). Patients were divided in 2 groups: group I – 22 children under 1year (mean age 0.12 ± 0.24) and group II – 23 children above 1year (mean age 5.9 ± 4.6).
Methods: Clinical symptoms, ECG, X-ray, EchoCG, 24h Holter ECG and the presence of congenital heart defects (CHD) were assessed. End points were PM implantation and survival.
Results: The significant differences were found in clinical presentation between the groups.

Symptom	Gr. I N = 22	Gr. II N = 23	p
Critical bradycardia	72.7%	78.3%	ns
Concomitant CHD	36.4%	8.7%	0.007
Congestive heart failure (CHF)	41%	0%	
Cardio-thoracic ratio (CTR)	0.6 ± 0.07	0.55 ± 0.06	0.028
Fractional shortening (FS)	38.2 ± 7%	44±5.5%	0.013
Adam-Stock attacks (MAS)	0%	43.4%	
Ventricular ectopy	34.8%	18.2%	0.04

Mitral regurgitation and wide QRS escape rhythm did not differ significantly between groups.
PM was implanted in 13/22 in Group I and 16/23 in Group II – all with critical bradycardia. Major indications for PM implantation in group I was CHF (89%) and LV congestion (66.7%) and in group II MAS (63.6%) and ventricular ectopy (40%)
The probability for freedom from PM implantation was 80%; 68.7%; 59.6%; 10% at 1; 5; 10 and 20 year of age.
Type of implanted PM was VVI with epimyocardial lead for all Group I patients, while in Group II it was physiologic (VDD or DDD) in 60% with endocardial leads in 91%.
Five children died – 4 from Group I and 1 from Group II. Probability of survival at 5, 10, 15 and 19 year of age is respectively 71%, 58.5%, 43.2%, and 25.4% and is significantly lower in Group I ($p = 0.0003$). PM related complications were present in 5 children, more frequent in Group I.
Concluisons: Clinical manifestations and indications for PM implantation are different in infants and older children. CCAVB has more favorable prognosis in the absence of CHD and age at diagnosis over 1 year.

P-96
Myocardial infarction and arterial thrombosis in homozygosity for the PAI-1 4G/5G polymorphism in identical newborn twins
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Myocardial infarction in children is rare overall in patients without congenital structural cardiac disease or Kawasaki disease. Reports of neonatal myocardial infarction and arterial thrombosis are extremely rare.
We report a case of monozygotic twins who both developed shortly after birth acute vascular events. They were delivered at 34 weeks of gestation by a healthy 25-year-old woman by caesarean section. The parents were of different ethnic groupsand they were unrelated. One neonate developed myocardial infarction (MI) due to thrombosis of the left coronary artery, the other thrombosis of the femoral artery. Both were treated with intravenous thrombolitic therapy. The twin with MI, on coronary angiography, showed left coronary anterior descendent thrombotic subocclusion.
Unfortunately, during cardiac catheterization, right coronary artery occlusion occurred with subsequent cardiac arrest. Intraaortic administration of urokinase was attempted to restore myocardial perfusion.
Both were discharged in good conditions. Antithrombin, protein C, protein S, lupus anticoagulants, homocysteine, factor V Leiden mutation, prothrombin 20210G→A polymorphism, methylenetetrahydrofolate reductase (MTHFR) 677C→T polymorphism and the −675 (4G/5G) polymorphism in plasminogen activator inhibitor 1 gene were analysed. Both twins were found to be homozygous for PAI-1 4G/5G polymorphism and H63D mutation (Hereditary haemochromatosis). The allele, PAI-1 4G/5G polymorphism, in association with intrapartum hypoxia and birth trauma probably contributed to increase the risk of arterial thromboembolisms in infants.

P-97
The molecular genetic basis of complex congenital heart defects – a study of 47 families
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Introduction: Congenital heart defects arise from interactions between environmental factors and genetic determinants.
Methods and results: The analysis comprised 35 families with more than one member suffering from the conotruncal heart defects (Group I) and 12 families (Group II) having a child with the clinical features of CATCH 22 syndrome. All family pedigrees were performed. Each patient was investigated by echocardiography to assess the diagnosis of the cardiac defect. Anamnestic information with regard to developmental millstones, learning abilities in childhood and psychiatric disorders were recorded. All individuals were qualified for further genetic molecular diagnostic procedures such as FISH analysis for microdeletion of chromosome 22q11 using probe N25 DiGeorge Region with 22qter control Direct CP 5141-DC. Based on the pedigree analysis in Group I we suggest that complex heart defects are transmitted as a recessive variant. None of the members of these families has the clinical features of CATCH 22 syndrome. In Group II we did not find the familial predisposition for the appearance of congenital heart defects. None of the evaluated members of the families from Group I had the microdeletion of chromosome 22q11 based on FISH analysis so we decided to isolate DNA for further molecular diagnosis.
In group II in 7 (60%) individuals with typical features for CATCH 22 syndrome FISH analysis confirmed microdeletion of chromosome 22q11.
Conclusions: 1. The huge progress in the molecular genetics creates new possibilities in the diagnosis of congenital heart defects.
2. The identification of the family with high risk of recurrence rate of conotruncal heart defects enable the genetic counselling and the high specialist medical care at the proper time.

P-98

Tree classification analysis predicts prognosis at presentation in infantile idiopathic dilated cardiomyopathy

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Background: idiopathic dilated cardiomyopathy (IDCM) has three possible follow-up at presentation: recovery, chronic heart failure, and death. Nevertheless, there is not any criterion to anticipate the prognosis.
Objective: select subgroups of children with IDCM by follow-up group using classification tree analysis.
Patients and Methods: this is a retrospective study of 179 children with IDCM (75 recoveries, 59 with chronic heart failure and 45 deceased). There were analyzed 36 clinical parameters and 25 from complementary exams. To build the tree it was employed CART algorithm, with selection by GINI index and prune by cost-complexity, aiming to maximize likelihood reason.
Results: From clinical and laboratorial data, it was built a tree with 10 branches and 9 nodes corresponding to seven variables, selecting 10 follow-up subgroups at presentation: two subgroups for death, five for chronic heart failure, and three for recovery. It was observed recovery in the subgroups: a) cardiothoracic ratio (CTR) less than 0.60; b) CTR from 0.60 to 0.70, functional class (FC) less than IV and age greater than 2.7 years old and c) CTR greater than 0.60, FC IV, mitral regurgitation (MR) 1–2, and age greater than 6.0 years old. Chronic heart failure was observed in the subgroups: a) FC less than IV and CTR greater than 0.70; b) FC less than IV, CTR from 0.60 to 0.70 and age less than 2.7 years; c) FC IV, CTR greater than 0.60, MR 1–2 and age less than 6 years old; d) FC IV, CTR greater than 0.60, MR 3–4, heart rate (HR) greater than 133 bpm and urea less than 23 mg% and e) FC IV, CTR greater than 0.60, MR 3–4, HR greater than 133 bpm, urea greater than 23 mg% and normal pediosous pulses. Deceased occurred in the subgroups: a) FC IV, CTR greater than 0.60, MR 3–4 and HR less than 133 bpm and b) FC IV, CTR greater than 0.60, HR greater than 133 bpm, urea greater than 23 mg% and small pediosous pulses.
Conclusion: From these select parameters, it is possible to anticipate follow-up category at presentation and adjust the best treatment to reduce the risk.

P-99

Predictors of supraventricular arrhythmia in adult patients with congenital heart diseases

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Introduction: Supraventricular arrhythmia (SVA) creates one of the main clinical problems in adults with congenital heart disease (CHD). Abnormal heart structure and performed cardiac surgery may cause arrhythmia which may leads to clinical deterioration.
Aim of the study: The assessment of risk factors for SVA in adults with CHD during long-term outcome.
Material and Methods: 1304 patients (P) (586 men) aged 18–72 (mean 29.4 ± 10), were studied and observated during 1–10 years (3.52 ± 1.83) (1995–2004). In P 25 different types of CHD were diagnosed – 13 simple: ASD, VSD, PDA, bicuspid aortic valve, pulmonary stenosis, subvalvular aortic stenosis, VAC, partial anomalous pulmonary venous connection, Marfan's syndrome, mitral insufficiency, idiopathic dilatation of the pulmonary trunk, additional vena cava superior sinistri, Wiliamas syndrome, 12 complex CHD: coarctation of the aorta, Fallot's tetralogy, ASDI, CAVC, Ebstein syndrome, univentricular heart, CCTGA, DTGA, DORV, pulmonary atresia, coronary fistula, Bland White Garland syndrom. SVA included: nonsustained supraventricular tachycardia (5xQRS HR > 100/min), atrial fibrillation/flutter. Anatomical complexity of the CHD, performed cardiac operation, hart failure (NYHA > I), cyanosis, age, gender were assessed during first visit. Kaplan-Meier curves were estimated, log-rank tests to compare curves, and Cox proportional hazards model for assessing multivariate associations between SVA and risk factors were calculated.
Results: SVA was observed in 10.3% of P. The probability of SVA occurrence after two years was 5.2% and 13.7% after 5 years. In univariate analysis the probability of SVA presence was higher in P: with complex vs simple CHD ($p = 0.00001$), operated vs unoperated ($p = 0.00001$), with NYHA > I vs NYHA I ($p = 0.00001$), cyanosis ($p = 0.00001$). Gender did not have the influence. The most predictive SVA risk factor in regression analysis was NYHA > I in all P (HR = 4.03), and in P with simple and complex CHD (HR = 4.21, HR = 4.94 respectively). Anatomical complexity (HR = 2.37), age in the whole population

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(HR = 1.34) and in P with simple CHD (HR = 1.46) were also significant risk factors. Operations decrease not significantly the relative risk of SVA occurrence. Cyanosis and gender did not demonstrate prognostic significance.
Conclusions: Risk factors for SVA are anatomical complexity, heart failure in all subgroups, age in all patients and in subgroup with simple CHD.

P-100
Electrocardiographic Features of Secundum Atrial Septal Defects with Pulmonary Hypertension in Adults
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Objectives: The development of pulmonary hypertension (PHT) is considered a risk factor in the long-term course of patients with atrial septal defect (ASD). The aim of this study was to evaluate the electrocardiographic features of secundum ASD with PHT, compared to those without PHT in adults.
Methods: Electrocardiography (ECG) was reviewed in 50 adults, mean age 34 ± 10 years (18–62 years) with secundum ASD who underwent cardiac catheterization. Electrocardiographic features were analyzed with reference to the right ventricular hypertrophy (RVH) criterion in adult. PHT was defined as mean pulmonary artery pressure (PAP) over 30 mmHg.
Results: The classical rsR' QRS morphology in lead V1 were in 30 patients and the other 20 patients showed Rs morphology (RS). RS was more frequent in ASD with PHT (Group 1, n = 20) than those without PHT (Group 2, n = 30) (p = 0.001). 9 patients with PHT showed small q wave before Rs, however, no one without PHT had qRs QRS morphology. In 20 patients with RS, height of R wave in V1 (V1R) over 5.5 mV (p < 0.01, 95% CI 0.808–1.05)) and R/S ratio in V1 (V1R/S) over 3.25 (p < 0.01, 95% CI 0.785–1.049) could discriminate the Group 1 from Group 2. In Group 1, V1R (r = 0.50, p < 0.05) and V1R/S (r = 0.56, p < 0.05) related with the degree of PAP. Among Group 1 patients, V1R over 10.5 mV (p < 0.01, 95% CI 0.98–1.00)) and R/S ratio in V1 (V1R/S) over 5 (p < 0.01, 95% CI 0.91–1.045) could discriminate the patients with PAP over 50 mmHg.
Conclusions: Most of ECGs in ASD patients with PHT showed the features of RVH instead of classical rsR' QRS morphology in lead V1. The indices of RVH criterion such as height of R wave and R/S ratio in V1 were helpful to differentiate the patients with PHT from those without PHT and further to discriminate the patients with severe PHT.

P-101
Echocardiogram and equilibrium radionuclide ventriculography in the evaluation of right ventricular dysfunction in patients with transposition of great arteries and Mustard procedure
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Objectives: To determine echocardiographic parameters predictable for right ventricle (RV) dysfunction evaluated with equilibrium radionuclide ventriculography in patients (pts) with systemic RV after Mustard procedure for transposition of great arteries (TGA).
Methods: we examined 38 pts (26 male; mean age 21 years) operated at 1 year or less with Mustard procedure for TGA. During 2006, during follow up visits, these pts were evaluated with equilibrium radionuclide ventriculography and echocardiogram M and B-Mode associated with tissue Doppler (TDI) of the tricuspid annulus.
Methods: Date are expressed as media (M) and standard deviation (SD). According to RV ejection fraction (RVEF) calculated with equilibrium radionuclide ventriculography we identify two groups of pts using 35% as cut-off (GROUP 1 = 16 pts with RVEF < 35%: M 26% DS ± 6.9%; GROUP 2 = 22 pts with RVEF > 35%: M 40.4% DS 3.9%). With echocardiogram we evaluated: tricuspid regurgitation (TR) (TR present in 7pts for GROUP 1 and 15 pts for GROUP 2), RV dilatation/RV DIL (RV DIL present in 11 pts for GROUP 1 and in 16 pts for GROUP 2), RV dysfunction/ RV DYSF (RV DYSF present in 5 pts for GROUP 1 and in 2 pts for GROUP 2), E wave at TDI (GROUP 1 M = 9.52 cm/ sec SD ± 2.5; GROUP 2 M = 10.58 cm /sec SD ± 1.5), A wave at TDI (GROUP 1 M = 5.98 cm/sec SD ± 3; GROUP 2 = 7.05 cm/sec SD ± 4.5), S wave (GROUP 1 M = 5.95 cm/sec SD ± 1.7; GROUP 2 = 8.8 cm/sec SD ± 2.7).
Results: Among echocardiographic parameters evaluated only S wave at TDI is statistically associated (t-test e-test ranksum <0.05) with a RVEF < 35% at equilibrium radionuclide ventriculography.
Conclusion: S wave at TDI is a good predictor of systolic disfunction of systemic RV and it can contribute in the evaluation of severity of RV dysfunction during follow up of pts with TGA and Mustard procedure.

P-102
Evaluation of sinus node function in young adults long term follow-up correction of d-transposition of great arteries by Senning atrial switch
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The aim of study: Evaluation of sinus node automaticity in asymptomatic young adults long term after reparation of d-transposition of great arteries repaired by Senning atrial switch.
Study population: We observed 12 pts (1 woman, 11 men) aged 18–21 yrs with d-transposition of great arteries after correction by Senning atrial switch, with electrocardiographic signs of sinus node dysfunction (SNA) and without history of syncope and/or symptomatic bradycardia.
All pts had episodic or sustain bradycardia in 12-lead and/or 24-hour ECG, 5 pts had an escape nodal rhythm, 2 of them had registered asymptomatic tachycardia-bradycardia syndrome. There were no RR pauses longer than 2.5 s.
Methods: All pts underwent transoesophageal atrial stimulation (RAS) for evaluation of extrinsic and intrinsic sinus node recovery times (SNRT), and corrected sinus node recovery times (CNRT), sino-atrial conduction time (SACT) and atrial refractory period (ERP-A). Pharmacological blockade (PHB) of sinus node was done with iv propranolol and atropine administration and intrinsic heart rate was assessed. SNRT > 1500 ms and CNRT > 525 ms were assumed as abnormal.
Results: Prolongation of extrinsic SNRT and/or CNRT with normalization after pharmacological blockade (functional SND) was observed in 8 pts (66.6%). The rest of patients presented normal values of sinus node recovery times. All patients reveals decreased intrinsic heart rate. Values of measured values are included in the table 1

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Table 1.

	HR	SNRT	CNRT	SACT	ERP-A
extrinsic	64.7	1582.4	698.7	128.5	280.0
intrinsic	76.7	1113.2	364.2	119.3	250.0

Conclusions:

1. Electrophysiological parameters of sinus node automaticity might be in normal range in spite of electrocardiographic signs of sinus node dysfunction in young adults with transposition of great arteries corrected by Senning atrial switch.
2. Both electrocardiographic monitoring and reduced intrinsic heart rate seems to be more useful than sinus node recovery times evaluation in assessment of sinus node dysfunction in this group of patients.

P-103

Physical activity in children operated for congenital heart disease

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Objectives: To assess the amount of physical activity and sports participation in children with completed surgical treatment for congenital heart disease (CHD) and in healthy controls.
Materials and Methods: Children 9–11 years old with completed surgical treatment for CHD and healthy age-matched, randomly selected controls performed seven days activity monitoring and were interviewed about their sports participation. Patients represented a cross section of all patients operated for CHD at our institution. Patients with surgery anticipated within a year were excluded, as were patients with chromosomal aberrations and/or neurological dysfunction. The activity monitor, ActiReg, records body position and movement of the trunk and legs, and assesses physical activity and energy expenditure.
Results: Complete results have been collected for 33 patients (15 boys and 18 girls) and 65 healthy controls (32 boys and 33 girls). Healthy boys spent more time in moderate-to-high physical activity (MHPA) than healthy girls, 64 (26) and 44 (16) minutes per day ($P < 0.001$). This gender difference was also found in patients, 58 (33) and 29 (15) minutes per day ($P < 0.01$). Healthy girls spent significantly more time in MHPA than patient girls ($P < 0.01$), but such a difference was not seen in boys. Healthy girls reached 60 minutes of MHPA 2.4 (1.9) days per week compared to 1.3 (1.7) days per week in patient girls ($P < 0.05$). Corresponding values for boys were 4.3 (2.0) and 3.4 (2.2) days per week (NS). Healthy girls had significantly more high-intensity sport participation compared to patient ($P < 0.05$). Such a difference was not found in boys.
Conclusions: Healthy girls exerted more physical activity than girls with completed surgical treatment for CHD, but in boys such a difference was not found. A contributing factor to the difference in girls could be more high-intensity sport participation among healthy girls. In all children there was a large discrepancy between the recommendation of 60 minutes of MHPA daily and the much lower recorded time in MHPA. The results stress the need of intervention for stimulating increased physical activity both in children with completed surgical treatment for CHD and in healthy children.

P-104

Neurodevelopmental and neuroradiologic outcome of UVH patients at age 5 to 7 years related to risk factor analysis

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Objective: Despite improved survival and neurodevelopmental outcome, children with hypoplastic left heart syndrome (HLHS) and other forms of univentricular heart (UVH) are still at increased risk for having cognitive, motor, and other neurological deficits.
Methods: We examined 27 children with HLHS or other forms of UVH at the median age of 5.70 (range 4.99–7.51) years and performed brain CT or MR imaging on 20. Possible risk factors were correlated with outcome.
Results: Mean full-scale intelligence quotient (FIQ) among the HLHS patients was 86.7, and among the other UVH patients 89.1, both differing significantly from the expected population mean ($p = 0.015$ and 0.029, respectively). Cerebral palsy was diagnosed in 1 of 7 HLHS patients and 2 of 20 UVH patients. Brain CT or MRI revealed ischemic changes and infarcts or atrophy in 5 of 8 patients who had undergone the Norwood procedure and in 2 of 12 of those who had not ($p = 0.062$). Abnormal CT findings correlated significantly with lower FIQ ($p = 0.045$) and verbal intelligence quotient (VIQ) values ($p = 0.02$). In the multiple linear regression model, the third post Rinkeli1 operative day diuresis after the primary operation and cardiopulmonary bypass time in the bidirectional Glenn (BDG) operation, correlated significantly with the primary outcome FIQ. Conclusions: In children with UVH, intellectual and neurologic deficits are common. Peri- and post-operative risk factors related to the primary phase and BDG operations contribute to these deficits

P-105

Natriuretic peptides within adults with secundum atrial septal defect (ASD II)

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Aim: The aim was to measure serum concentrations of C-terminal ANP and BNP in adults with ASD II successfully closed with Amplatzer Septal Occluder within 12-month follow-up and to correlate them with transthoracic echocardiographic (TTE) parameters.
Materials and Methods: 39 consecutive patients with ASD II enrolled a study group. 4 patients have withdrawn the consent after one month. There were 29 (74%) women and 10 men in the age between 19 and 66 years. The mean age was 39 ± 13 years. All subjects had baseline, 1, 6 and 12 month checkups contained of TTE, ANP and BNP evaluation. Qp:Qs ratio ranged from 1.5 to 4.3, mean value 2.2 ± 0.8. There was a control group consisted with 16 patients who had TTE and ANP and BNP evaluation. p values <0.05 were considered to be statistically significant.

Results: Table 1 shows ANP and BNP values within one year follow-up.

	Time			
NP type	Baseline	1 month	6 months	12 months
ANP (pg/ml)	36.0 ± 19.7	36.6 ± 24.1	33.3 ± 17.1	32.5 ± 17.4
BNP (pg/ml)	27.8 ± 32.0	43.1 ± 37.6	30.1 ± 27.5	27.2 ± 21.9

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Table 2 shows ANP and BNP below and over 40 year of age.

	Age < 40 years	Age ≥ 40 years	p
ANP (pg/ml)	26.8 ± 14.5	41.1 ± 23.7	0.05
BNP (pg/ml)	13.7 ± 12.4	37.7 ± 26.8	0.001

Table 3 shows ANP and BNP in patients without and with systolic pulmonary hypertension.

	RVSP < 35 mmHg	RVSP≥ 35 mmHg	*p*
ANP (pg/ml)	25.9 ± 16.1	43.1 ± 23.5	0.036
BNP (pg/ml)	11.0 ± 10.3	41.8 ± 25.5	0.0001

Table 4 shows baseline TTE parameters correlations with BNP.

Parameter TTE	P	R
RALSd	0.000005	0.675
RADa	0.003	0.476
Qp:Qs	0.03	0.351
RVSP	0.04	0.393

Table 5 shows TTE parameters correlations with ANP

Parameter TTE	*p*	R
MPAd	0.006	0.457
RALSd	0.02	0.380
Qp:Qs	0.02	0.393

Conclusions:

1. ANP and BNP concentrations reflect an interatrial shunt in adults with ASD II.
2. Patients with ASD II demonstrate higher ANP and BNP concentrations than control group.
3. RALSd, being a morphological parameter, the best reflects significance of a shunt within the study population.
4. Patients with systolic pulmonary hypertension present with higher BNP and ANP values.
5. Patients over 40 year of age present with higher BNP values.
6. BNP one month after was significantly higher than before the procedure probably due to LVDd enlargement.
7. ANP and BNP values 6 and 12 months after the procedure did not differ from the baseline ones.

P-106

Patients with aortic coartation seen at a GUCH outpatient clinic

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Objectives: Assessment of the characteristics and mid-term outcome of patients with aortic coartation seen at a GUCH outpatient clinic.
Methods: Retrospective descriptive analysis of 128 patients older than 16 years who had a follow-up visit within a two year period (2005–2006). The median follow-up time: 22.3 years. Mean age: 25 years.
Results: The median of diagnosis age was 46 days and 114 patients (89%) underwent surgical repair of their coartation a mean age of 329 days. Coartaction site was: 86% yuxtaductal, 27% postductal and 13% preductal. Associated congenital cardiac structural anomalies were found at diagnosis time on 74% of the patients (21% ductus arteriosus; 19% ventricular septal defect; 18% hypoplastic aortic arch). Presenting diagnostic findings were: cardiac murmur (45%), congestive heart failure (41%), systemic hypertension (3.9%) and absent femoral pulses (4%). Operative techniques included: end-to-end anastomosis (45%), subclavian flap aortoplasty (39%) and Dacron-patch aortoplasty (1%). Twenty-six patients (20%) underwent a redo procedure: balloon dilatation (48%) and Dacron-patch aortoplasty (35%), at a mean age of 13.37 years and a mean interval after the original repair of 13.39 years. Six patients needed a third intervention: balloon dilatation (2 cases) carotid-to-thoracic aorta bypass grafting (2 cases), stenting (1 case) and end-to-end anastomosis (1 case) at a mean age of 17.69 years (mean interval after the second procedure of 9.75 years). Mean blood pressure at the last follow-up visit was 125/52. Eleven patients were found to have a dilated ascending aorta, which bore a definitive association with a bicuspid aortic valve, the latter having a prevalence of 44% of the cases. Late cardiovascular complications were: recoartation (20.5%), systemic hypertension (12.7%), aortic aneurysm (7%), subclavian steal 0.8%), whereas neither intracranial bleeding nor endocarditis were encountered. Nine pregnancies have been closely controlled, with an uneventful course.
Conclusions: Our patient's features do not differ from other series. Mid-term outcome of repaired aortic coartation is excellent, notwithstanding all patients should be follow-up on a regular and long-term basis

P-107

Carotid-Subclavian Artery Index: Validation of an Echocardiographic Index to detect Coarctation

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Introduction: Due to patency of the arterial duct and the parallel circulation during the fetal life, coarctation remains a difficult diagnosis prenatally and even shortly after birth. Milder forms of coarctation can be missed until the child present with sudden collapse after closure of the ductus arteriosus. Firstly, our study aimed to assess accuracy of a new cardiographic index based on morphologic measurements of the distal aortic arch, the Carotid-Subclavian Artery Index (CSA Index), in detecting coarctation in newborns, infants and children, independently of the presence of a patent ductus arteriosus and other cardiac lesions. Secondly, to assess the additive value of another morphologic index in predicting coarctation, the I/D ratio, based on the measurement of the aortic isthmus and the descending aorta.
Methods: It is a retrospective cohort study in a tertiary care children's hospital. Echocardiograms recorded from children aged 0 to 18 years between 1996 and 2006 with a coarctation were reviewed. Offline echocardiographic measurements of great vessels and aortic arch dimensions were done for each participant to calculate their CSA Index and I/D Ratio. CSA index is the ratio of the distal transverse aortic arch diameter (DTA), to the distance between the left carotid artery and the left subclavian artery (LCSA). I/D ratio is the ratio of isthmus (I), to descending aorta diameter (D) (Fig. 1A). Values of CSA Index and I/D ratio from coarctation group were compared with those from a normal local control population.
Results: 69 echocardiograms from patients with coarctation were analysed. Compared with controls, patients with coarctation

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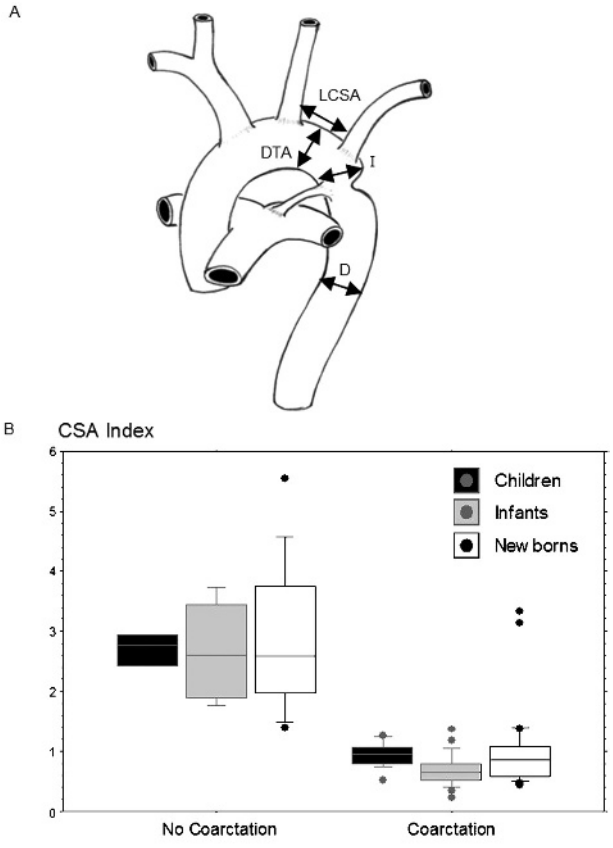
had a significantly lower CSA index (0.88 ± 0.49 vs 2.75 ± 1.04, $p < 0.0001$) and I/D ratio. The same significant difference was observed, independently of age (Fig. 1B) and other associated defects, even complex ones. CSA Index confirmed its good sensitivity and specificity (97.1% and 92.3% respectively). This was not improved by adding the I/D ratio.



Conclusions: An abnormal CSA index is highly suggestive of coarctation independently of age, of the presence of a patent ductus arteriosus or of other cardiac defects. The addition of another anatomic index, the I/D ratio, was not helpful in our study.

P-108
Quantitative Assessment of Right Ventricular Function Using Three Dimensional SSFP Magnetic Resonance Angiography

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Introduction: So far the gold standard for right ventricular (RV) stroke volume (SV) measurement has been two-dimensional (2D) cine magnetic resonance imaging (MRI). Endsystolic and enddiastolic volumes were assessed by manually tracing endocardial borders of the RV in a stack of slices perpendicular to the long axis of the RV. Major problems were patients unable to cooperate with breath-holding, distinction between the atrial and ventricular lumen in basal slices during the cardiac cycle, time consuming manual tracing of RV trabeculations and slice thickness usually greater than 5 mm. In this study a new three-dimensional (3D) method for volumetric RV-SV assessment was evaluated.
Methods: The whole heart (n = 18, mean age 21.7, 15.2 to 28.8 years) of healthy volunteers (n = 13) and patients with competent atrio-ventricular valves (tetralogy of Fallot n = 3, aortic coarctation n = 2) was imaged with a free-breathing vector-ECG triggered and navigator gated 3D steady-state-free-precession (SSFP) sequence with a T2-preparation pulse on a commercial 1.5 T MRI scanner (Philips Medical Systems, Best, Netherlands) during endsystole and enddiastole (flip angle of 90°, 1.0 mm^3 isotropic voxels, TR 3.6 ms, TE 1.8 ms). Using a simplex-mesh model the endocardial borders of the RV were traced semiautomatically. The planes of the pulmonary and tricuspid valve were defined in the 3D image data set on multiplanar reformatted slices. Papillary muscles within the RV were excluded defining a threshold separating blood and myocardium. Results were compared to 2D-SSFP-Cine-MRI (6 mm continuous slices, 25 phases) and flow measurements across the pulmonary artery (PA). Pearson's correlations and Bland and Altman tests were performed.
Results: Pearson's correlations are summarized in Table 1 with highest correlations for 3D-SSFP excluding trabeculations from the RV-volume regarding intra- and interobserver variability and comparing these results with PA flow measurements. Comparing RV SV excluding papillary muscles with flow across the PA the mean difference was −1.7 ml ± 7.3 ml (Bland-Altman test).
Conclusions: Volumetric RV analysis of 3D SSFP datasets demonstrates high correlation with PA-flow as well as low intra- and interobserver variability. As these datasets allow highly accurate volumetric analysis of RV function in free-breathing subjects this method may be very helpful in sedated patients with abnormally shaped right ventricles.

Table 1

Pearson's correlation (p = 0.01)	SV-RV (3D-SSFP) (including trabeculations)	SV-RV (3D-SSFP) (excluding trabeculations)	2D-Cine- SSFP
Intra-observer	0.91	0.94	0.87
Inter-observer	0.86	0.93	0.72
Flow across PA	0.78	0.89	0.79

P-109
Does cardiac magnetic resonance imaging really replace diagnostic catheterization in congenital heart disease?

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Objectives: To determine the real influence of cardiac MRI on the number and indications of diagnostic catheterization procedures in congenital heart disease.
Methods: Retrospective analysis of number and indications of diagnostic and interventional catheterization procedures compaired to cardiac MRI-studies between 1999 and 2005 in a pediatric cardiology tertiary referral center.
Results: Triplication of cardiac MRI-studies (1999: 80, 2005: 269). Decline in diagnostic catheterization procedures by 66% (1999: 390, 2005: 134). 41% (111 of 269) cardiac MRI-studies really replacing

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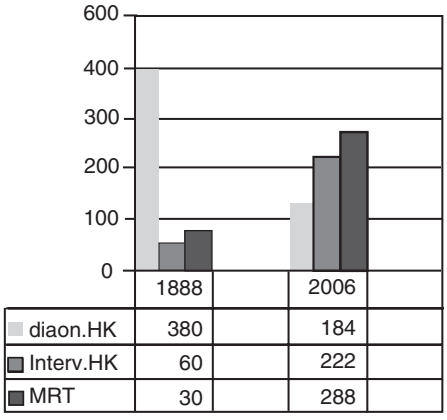
neccessary catheterizations in 2005. 13% (34/269) of MRI-studies preliminary to interventional caths. 22% (61/269) due to clinical indications like myocarditis and arrhythmia. 24% (63/269) were made during multi-center studies of right ventricular function in congenital heart disease.
Mean length of MRI-studies ("door-to-door time"): (1999: 83 minutes, 2005: 87 min., anaesthesia-studies: 108 min.



Conclusions: Diagnostic catheterization in congenital heart disease has decreased substantially in the last years. Cardiac MRI does replace a big part of catheterizations (43%) but is increasingly been used in planning interventional catheter-procedures and in the hemodynamical follow up after corrective heart surgery.

P-110

Non-invasive Assessment of Congenital Pulmonary Vein Stenosis in Children Using Cardiac-non gated CT with 64-slice Technology

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Objective: The aim of the study was to investigate the accuracy of cardiac-non gated CT using 64-slice technology in detecting congenital pulmonary vein stenosis in children.
Background: Management of congenital pulmonary vein stenosis is a diagnostic challenge. Echocardiography may be insufficient so that cardiac catheterization is the reference standard in this setting. New generation of multislice computed tomography may play an important role in the non-invasive evaluation of this anomaly.
Methods: Twelve CT examinations were consecutively performed from may 2005 to December 2006 in children aged 1.5–12 months (median 5 months) for suspected congenital pulmonary vein stenosis. Cardiac-non gated CT acquisitions were performed after peripheral injection of contrast agent using bolus tracking technique. Pulmonary veins were evaluated for their pattern of connectivity from the lung to the left atrium and for the presence of stenosis or atresia. CT findings of pulmonary vein stenosis were compared with combined findings available from echocardiography, catheterization, and surgery.
Results: Pulmonary venous drainage from the right lung (n = 27) and left lung (n = 24) were evaluated as separate structures (n = 51). Of the 51 structures 22 had surgical and/or catheterization data and 41 had echocardiography for comparison. CT visualized 100% (41/51) of the investigated structures, while echocardiography visualized 80% (41/51). Among the 12 examinations CT identified 23 stenotic or hypoplastic pulmonary veins. CT agreed with echocardiography in 38% (8/23) and surgery/catheterization in 100% (22/22) of the available comparisons.
Conclusions: Cardiac-non gated pulmonary venography assessed the pulmonary veins more completely than echocardiography. It is a reliable method of detecting pulmonary venous stenosis in children. Cardiac-non gated CT may serve as a non-invasive alternative imaging modality in this setting.

P-111

Mitral Valve Annular Function in Children with Normal and Regurgitant Valves: A Three Dimensional Echocardiographic Study

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Introduction: The mitral valve is a complex structure, the true shape and function of which are difficult to visualise accurately in only two dimensions. Our aim was to characterise mitral valve (MV) annular motion in normal children's hearts and in those with mitral regurgitation (MR), using Real Time Three Dimensional Echocardiography (RT3DE).
Methods: RT3DE full volume loop datasets, acquired in consecutive subjects with mitral regurgitation (n = 17) and age matched controls (n = 20), were analysed offline. Simultaneous multiplanar review was used to delineate and measure MV annular areas in systole and diastole.
Results: MV annulus area decreased in diastole and increased in systole in both groups. The annulus in patients with MR is dilated when compared to normal subjects, being significantly larger in diastole (systolic mean 6.65+2.21 cm^2/m^2 in patients with MR and 5.28 cm^2/m^2+1.68 in normal subjects, p = 0.091, diastolic mean 5.13+1.63 cm^2/m^2 in patients with MR and 3.05 cm^2/m^2+0.90 in normal subjects, p < 0.0001). The percentage change in MV annulus area from systole to diastole showed a trend towards being smaller in patients with MR, although this did not reach significance (21.36% vs 41.13%, p = 0.189). A sub-group analysis of patients with moderate or severe MR showed MV excursion, expressed as a percentage of LV length, to be significantly less than in normal subjects (14.23+6.40% vs 17.04+3.16%, p = 0.036).
Conclusions: In contrast to adults, MV annular area in children decreases in diastole and increases in systole. In children with mitral regurgitation, the annulus is dilated and the dynamic annular function is depressed.

P-112

Intraoperative evaluation of a MicroMultiplane Transesophageal Echocardiographic Probe in Surgery for Congenital Heart Disease

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Introduction: The superior imaging performance of transesophageal transducers for multiplane Doppler echocardiography has been demonstrated in pediatric patients undergoing cardiac surgery. To date, however, the size of these probes has limited their use in neonates and small children. New technologies that enable to perform transesophageal echocardiography (TEE) in smaller patients are therefore important.
Methods: Clinical experience was acquired in 20 patients using the Philips MicroMultiplane TEE probe (7–8.2 mm diameter tip with

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a 5.2 mm diameter shaft) which was specifically designed for use in neonates weighing as little as circa 2 kg.
Results: Patients were examined intra-operatively using the TEE probe. The examined patients ranged in age from 8 days–9 months and in weight from 2.5–6 kg. Five of these 20 patients weighed less than 3 kg. There was a large variation in diagnoses of which the most prevalent were tetralogy of Fallot (n = 7), hypoplastic left heart syndrome (n = 2) and (atrio)ventricular septal defect (n = 6). In all patients it was possible to get adequate echoviews. In 2 patients we had to adapt the ventilatory settings because of increased airway resistance after probe insertion. In 2 patients surgical re-intervention was performed due to TEE assessment immediately after weaning from bypass; in 1 patient significant obstruction of the right ventricular outflow tract was still present after Fallot correction; and in 1 patient an additional muscular ventricular septal defect was still present after ventricular septal defect closure. This reintervention rate is comparable with the 5–10% reported earlier in pediatric cardiac surgery after TEE assessment.
Conclusions: The MicroMultiplane TEE probe provided excellent diagnostic intra-operative TEE in neonates and small children without major complications. However, special attention should be paid to the ventilatory parameters in neonates weighing less than 3 kg.

P-113
Assessment of coronary artery aneurysms in patients with Kawasaki disease by multislice spiral computed tomography in follow-up

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Background: Multislice spiral computed tomography (MSCT) is used as non-invasive methods for visualization of coronary arteries (CA) in adults. The experience in children is limited.
Objective: To assess the feasibility of MSCT in coronary artery aneurysms in children with Kawasaki disease (KD) in follow-up.
Methods: The 16-slice MSCT scanner was used. A bolus injection of nonionic contrast medium was given. All images were reconstructed by the segmental reconstruction methods. Each examination was performed during breathholding. All patients were under short general anesthesia. Heart rate range 50 to 100 /´(median – 91.5). A beta-blocking agent was used in 2 cases. All patient underwent transthoracic echocardiography (TTE) at the same time.
Material: Eight patient (7 boys, 1 girl) age from 21 to 184 months (median – 109) with history of Kawasaki disease complicated by coronary artery aneurysms (CAA) were enrolled. In acute stage 4 patients had medium CAA and 4 patients had giant CAA. The follow-up period was 13 to 148 months (median – 67).
Results: Coronary artery imaging by MSCT was feasible in all patients. Scan time was 15–22 seconds, entire examination was generally completed within 15 minutes. Among 4 patients with medium CAA in acute stage: aneurysm persists; in 1pt in left CA, in 1 pt in right CA with calcification; in 1pt right CA was occluded with calcification in both coronary arteries. In 1 case right and left CA were normal. Among 4 patients with giant CAA in acute stage: in 1 pt very narrow both coronary arteries with calcification; in 1 pt giant CAA in left CA with calcification and right CA occluded with calcification; in 2 pts giant CAA in left CA were observed. There were no adverse reactions after nonionic contract medium and short general anesthesia. In TTE proximal segments of CA were seen; occlusion of right CA and calcification was not detected.
Conclusions: 1. MSCT is useful non-invasive method to assess the progression of changes in coronary arteries in KD 2. MSCT in short general anesthesia is safe 3. MSCT has may become a standard diagnostic tool in patients with CAA in Kawasaki disease.

P-114
Tissue Doppler imaging analysis of right ventricular function before and after transcatheter closure of atrial septal defect

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Introduction: Patients with an atrial septal defect (ASD) have long-standing right ventricular dilation and dysfunction. We used Tissue Doppler Imaging (TDI) to assess right ventricular systolic and diastolic function in patients with ASD before and after closure with the Amplatzer septal occluder.
Methods: 52 pts with ASD secundum, mean age 25yrs (5–63), 19 male and 33 female underwent transcatheter closure of their ASD with an Amplatzer septal occluder. ASD size was 8–34 mm by echocardiography and mean Amplatzer size was 22mm (8–40). After conventional echocardiographic assessment, pulsed TDI was obtained from the apical 4-chamber view at the basal RV free-wall-tricuspid annular junction. The following measurements were made: peak early systolic myocardial tissue velocity (e), peak late systolic myocardial tissue velocity (a), e/a ratio, peak systolic myocardial tissue velocity (s). Tricuspid annular motion was measured by M-Mode echocardiography in the 4-chamber view.

Results	Pre-Amplatzer	Post-Amplatzer	p
E (cm/sec)	19 ± 2	16 ± 2	<0.01
A (cm/sec)	15 ± 4	11 ± 4	0.05
E/A	1.37 ± .25	1.45 ± 2	NS
S (cm/sec)	18 ± 2	16 ± 2	<0.01
TV M-Mode (mm)	33.7 ± 6	25.2 ± 3	<0.02

Conclusion: There is dramatic improvement in both systolic and diastolic right ventricular function, measured by TDI indices the next day after transcatheter ASD closure, even in pts with long-standing RV dysfunction. Very high myocardial tissue velocities tend to normalize soon after the device placement. This is probably due to the sudden relief from the long-standing volume overload of the right ventricle secondary to the abrupt removal of the left to right shunt.

P-115
QT and QTc intervals dispersion in children with chronic renal failure

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Objectives: The study of QT and QTc dispersion in children with chronic renal failure (CRF) treated with chronic haemodialysis.
Methods. Patients: 15 patients, aged between 12–18 years, with CRF in end stage renal disease (ESRD), treated with haemodialysis in 3 sessions/week. The surface electrocardiogram to manually measure on three successive cardiac cycles the values of QT/QTc intervals, QT dispersion (QTD) and QTc dispersion (QTcD) (Bazett's formula) was performed before the dialysis session.

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These values of QTD and QTcD were compared to similar values from 20 healthy children, without any history of renal and/or cardiovascular disease and also from 10 children with acute postinfectious glomerulonephritis.

Results: The QT and QTc interval dispersion values were higher than those in normal children (QTD: 40 ± 18.56 ms; QTcD: 55.47 ± 20.68 MS) in 10/15 cases in patients with chronic renal failure treated with chronic haemodialysis (QTD: 59.67 ± 21.32 msec and QTc D: 77.25 ± 44 msec) and in 4/10 cases with acute postinfectious glomerulonephritis (QTD: 46.86 ± 20.3 msec, and QTcD: 71.58 ± 31.22 msec). Even if QTD and QTcD are higher in cases with acute postinfectious glomerulonephritis than in normal subjects, they are significantly lower than those in children with chronic renal failure. We could not establish any correlation between the increased QTD/QTcD and the biochemical changes during the haemodialysis session.

Conclusions: The significant increasing of QTD and especially QTcD in children in ESRD, hemodialysed, constitute an useful marker for the risk of the severe ventricular arrhythmias which can occur among these patients and justifies the systematic search for this parameter ECG.

P-116

Increased torsion, measured by Magnetic-Resonance-Tagging, of both ventricles in patients with Atrial Septal Defects normalises after interventional occlusion of the defect

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Introduction: It is unclear how right and left ventricular contraction are affected by interventional Atrial Septal Defect (ASD) occlusion. Magnetic-Resonance-Tagging (MR-Tagging) is a validated method for point to point contraction analysis of the ventricles. The aim of this study was to determine right and left ventricular torsion and circumferential strain in patients before and after ASD occlusion.

Patients and Methods: Ten patients (28 ± 25 yrs) before and after Amplatzer ASD occlusion and eight normal subjects (age 25 ± 16 yrs) were studied by MR-Tagging.

By MR-Tagging in short axis at three different levels torsion of both ventricles was measured during systole and diastole. Systolic and diastolic torsion were summed up and given in degrees of total ventricular torsion. Circumferential strain was measured according to $CS(hp) = [1 - CL(hp)/CL(0)]$, where CS(hp) is circumferential strain at a given heart phase and CL the length of calculated center line in the myocardium.

Volumes of both ventricles were measured by conventional steady-state-free-precession scans. Left-to-right shunt was measured by MR flow measurements in the main pulmonary artery and ascending aorta (Qp/Qs).

Results: Patients before ASD occlusion had increased torsion in both ventricles compared to normal subjects (RV at base: 7 ± 3° vs 4 ± 1°, $p < 0.05$ and LV at base: 10 ± 4° vs 6 ± 1°, $p < 0.05$). After ASD occlusion torsion in both ventricles normalised. Torsion and shunt volume had a negative linear correlation ($r^2 = 0.86$; $p < 0.01$).

Patients before ASD occlusion had decreased circumferential strain in the right ventricle compared to normal subjects (81 ± 3 vs 84 ± 1%, $p < 0.01$). After ASD occlusion circumferential strain in the right ventricle normalised. Circumferential strain in the left ventricle was not significantly different before and after ASD occlusion and compared to normal subjects.

After ASD occlusion Qp/Qs normalised from 2.0 ± 0.5 to 1.0 ± 0.3 ($p < 0.05$). The right ventricular enddiastolic volume decreased from 102 ± 29 ml/m² to 88 ± 18 ml/m² ($p < 0.05$). Left ventricular enddiastolic volume did not change significantly from 58 ± 15 ml/m² to 71 ± 18 ml/m².

Conclusion: Right ventricular volume load due to ASD increases total ventricular torsion in both ventricles. After ASD occlusion torsion of both ventricles normalises.

P-117

Interventional treatment in postoperative pulmonary stenosis and hypoplasia in children with HLHS during multistage operative treatment

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Background: Success of multistage treatment in HLHS patients is based on many hemodynamic aspects. The most important of them are pulmonary pressure, resistance and pulmonary arteries diameter. Hypoplasia and stenosis of pulmonary arteries after Norwood operation are connected with changes in geometry of pulmonary arteries and significantly deteriorate prognosis in this group of patients.

Aim: The aim of this study was the estimation of interventional treatment efficiency in pulmonary arteries stenosis and hypoplasia in HLHS patients after second stage of palliative treatment – bidirectional Glenn anastomosis.

Materials and Methods: Between year 1999 and 2006 we analyzed 21 patients with HLHS and postoperative pulmonary stenosis or hypoplasia aged 6 months to 6 years. Total number of interventions in this group was 31. In 6 patients with narrowed left pulmonary artery we performed 7 balloon angioplasties. In 15 patients with pulmonary hypoplasia we performed balloon angioplasty with stent implantation during the same procedure (8 pts); and with delayed stent implantation in 7 pts. Three patients required balloon stent redilatation after 6 months to 2 years after primary stent implantation. In 1 patient stent was implanted intraoperatively during bidirectional Glenn anastomosis. Mean pulmonary pressure and diameter of right and left pulmonary arteries were estimated directly after intervention.

Results: Balloon angioplasty was efficient in only 4 patients with pulmonary stenosis and totally ineffective in the group of children with pulmonary hypoplasia. In this group (15 patients) primary or delayed stent implantation resulted in efficient diameter increase from 150 to 300% comparing with primary size of vessel. We also succeeded in significant mean pulmonary pressure reduction (mean 4.5 mmHg) ($p = 0.04$).

Conclusions: 1. In children with HLHS and coexistent post-operative pulmonary arteries stenosis transcutaneus balloon angioplasty and stent implantation is an efficient method of treatment resulting in vessel diameter increase and pulmonary pressure reduction.

2. Procedures of isolated balloon angioplasty are efficient only in patients with pulmonary stenosis.

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P-118

Transcatheter Recanalization of Acutely Occluded Aortopulmonary Shunts

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Objective: To describe the efficacy of balloon dilation for recanalization of acutely occluded aortopulmonary shunts.
Patients and Methods: Seven infants (age 5 days to 7 months) with shunt-dependent pulmonary blood flow and clinical evidence of acute shunt occlusion underwent emergency cardiac catheterization, with a view to reopening the shunt (central shunt n = 6; modified Blalock-Taussig shunt (n = 1)). The interval from surgery to cardiac catheterization was 1–45 days. Two patients had aortopulmonary collateral vessels as an alternative source of pulmonary perfusion; 3 had anterograde flow from the right ventricle, and 3 infants had a small duct which had not been ligated at the time of shunt surgery. The shunts could be crossed with an 0.018" Terumo wire and 4F end-hole catheter combination. Balloon angioplasty was performed using a balloon with the same nominal diameter as that of the shunt (4 mm or 5 mm).
Results: All shunts were successfully recanalized. Two patients with recurrent severe cyanotic episodes despite documented shunt patency underwent stent implantation in the right ventricular outflow tract at the same procedure (the shunts were patent at repeat cardiac catheterization 3 and 6 months later respectively). One infant required shunt revision 24 hours later due to intermittent changes in systemic saturation; a small clot was seen at the distal anastomosis. The remaining patients required no further procedures upto a follow-up of 6 months, or earlier definitive repair. None of the patients has had documented neurologic sequelae resulting from the hypoxemic insult.
Conclusions: Balloon angioplasty is feasible and effective in recanalizing acutely occluded shunts, without the need for further immediate surgery in the majority of patients.

P-119

Stenting of the transverse arch in arch hypoplasia after surgical repair of coarctation of the aorta

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Introduction: Coarctation of the aorta (CoA) is often correlated with various forms of arch hypoplasia. Surgical repair in infancy by lateral thoracotomy and extended resection or patch augmentation is currently the treatment of choice in simple CoA and mild forms of arch hypoplasia. When residual arch hypoplasia reveals during follow-up, complex redo-surgery with arch augmentation by using deep hypothermic circulatory arrest may be indicated. This commonly is correlated to significant perioperative morbidity and possibly mortality. We hypothesised that interventional treatment may be feasible in this selected patient group.
Methods: Prospective investigation over a 6 month period in all patients presenting with significant arch hypoplasia after CoA repair. Morphologic evaluation by MRI and physiologic assessment by using a catheter based pharmacologic stress protocol using a bolus administration of Orciprenaline. Additional factors evaluated were the minimal diameter of the re-CoA site and the diameter of the aorta at the level of the diaphragm (dAo). Subsequent stent implantation was performed and the pharmacologic stress testing was repeated immediately thereafter.
Results: A total of 5 patients was detected. The minimal arch diameter was 9.80 mm (SD 3.63 mm) or 59% of dAo (19.00 mm; SD 3.32 mm). The gradient at rest was 23.40 mmHg (SD 8.76 mmHg), the gradient after the orciprenaline bolus increased to 82.00 mmHg (SD 18.23 mmHg). Stent-Implantation (short CP-Stents mounted on short balloons) was feasible in all patients without impairment of the carotid arteries. The arch diameter increased to 16.40 mm (SD 3.29 mm) or 87% of dAo. There was no residual gradient at rest and after a second bolus of Orciprenaline 9.80 mmHg (SD 3.63 mmHg).
Conclusions: Stenting of the transverse arch seems feasible in patients after CoA-repair. Good haemodynamic results at rest and under pharmacologic stress testing can be achieved by using a diameter of about 90% of dAo. This technique offers an attractive alternative to complex surgical intervention.

P-120

Dilatation of aortic coarctation in infantswith severe left ventricular dysfunction: a bridge to surgery

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Background: Dilatation of neonatal aortic coarctation is controversial and elective surgery is preferred in most centres even in the presence of ventricular dysfunction. The aim of our study was to evaluate the results of emergency balloon dilatation in neonates with aortic coarctation and severe left ventricular dysfunction.
Patients and methods: From June 2004 to June 2006, 15 neonates with aortic coarctation and severe ventricular dysfunction underwent emergency balloon dilatation. Diagnosis were: simple coarctation (9), coarctation and CIV (3), coarctation and aortic stenosis (2), complex TGA (1). Mitral incompetence was present in 11, metabolic acidosis in 8, severe pulmonary hypertension in 9. Eleven infants needed mechanical ventilation, all were under PGE1.
Results: In all but 2 neonates it was possible to dilate the aortic isthmus. Early complications included cardiac arrest in 4 patients and localized dissection in 2. There were 3 peri-procedural deaths and 3 more deaths due to persistent left ventricular dysfunction (1 post-surgical) (40%). In 9 patients femoral pulses reappeared immediately after dilatation and metabolic acidosis regressed. Eight out of nine survivors underwent elective surgery after a mean period of 10 days (1–60) from angioplasty. Two infants with localized coarctation and a good immediate result have not been operated so far. At a median follow-up of 14 months, all survivors have a normal left ventricular function and normal blood pressure.
Conclusions: Aortic coarctation in neonates with ventricular dysfunction has a severe prognosis. Emergency dilatation offers a good palliation and can constitute a bridge to surgery in very ill patients.

P-121

Pulmonary stent implantation in children with single ventricle before and after completion of cavopulmonary connection

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Pulmonary artery stenosis and hypoplasia are risk factors in patients with single ventricle and staged palliation of cavopulmo-

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nary connection (CPC). We report our results on 11 children with functional univentricular heart who received pulmonary stent implantation in our institute since January 2002.
Patients: All 11 patients had complex congenital heart disease, 8 with functional single right, 3 with a single left ventricle. Patients' age ranged from 0.1 to 16.5 years (mean 6.4 years) and body weight from 2.4 to 63 kg (mean 22.7 kg). Within the process of staged palliation of CPC 2 patients were treated after aortopulmonary shunt implantation, 6 (55%) after Glenn procedure and 3 after CPC completion. Mean interval after preceding surgery was 44.2 (1–132) months. Indication for stent implantation was a morphologic stenosis with a pressure gradient, and/or hypoplastic pulmonary vessels behind the stenosis, and a poor result after balloon dilatation alone.
Results: Interventional procedure could be performed in all cases successfully with satisfactory results and without any complications. Site of implantation was left pulmonary artery (LPA) (n = 7), right pulmonary artery (RPA) (n = 4) and Glenn anastomosis (n = 1). We implanted 5 Jo-Stents, 5 Palmaz Genesis stents, 1 coronary stent and 1 ev3 stent. During a mean follow-up of 29 (1 to 57) months 6 patients (55%) received their next surgical step of CPC palliation. Within this surgery 2 stents were removed and the vessel (LPA) reconstructed, 4 stents were left in place. In both patients with surgically removed stents the pulmonary stenosis reoccurred, in 1 patient even with a complete occlusion of the LPA, which caused a major hemodynamic problem. Both received a second stent at the same site.
Conclusion: This study demonstrates the efficacy and safety of percutaneous stent implantation to correct pulmonary artery stenosis in children with CPC palliation. LPA is mainly affected, often due to compression by the large ascending aorta. In a following surgical step of CPC palliation previously implanted stents at this site should be left in place or should be exchanged to a larger diameter in a hybrid procedure, because the cause of stenosis may not change after surgery.

P-122
Mid-long term follow-up results of stent dilatation of Aortic coarctation
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Objectives: To evaluate the mid-long term outcome of stent dilatation of aortic coarctation. focusing on the impact of the procedure on arterial blood pressure and on aortic wall complications.
Patients and methods: Twenty-four pts (mean age 21 ± 17, range 4–67 years, mean weight 53 ± 16, range 20–80 kg) with stent dilatation of native (14) or recurrent postoperative (10) aortic coarctation. Indications to dilatation were: Color Doppler Echo and/or MRI diagnosis of Coarctation associated with arm-to leg systolic pressure gradient (SPG) ≥20 mmHg and/or systemic hypertension. The follow-up protocol included physical examination, systolic blood pressure at right arm measurement (rSBP) and Color Doppler Echo at 1, 3, 6, 12 months and every year after the procedure. Exercise test, 24 hrs BP monitoring and CT scan or MRI were performed 6 months after the procedure.
Results: Stent dilatation was successful in all cases (mean hemodynamic SPG from 36.7 ± 22 to 1.8 ± 3.5 mmHg ($p < 0.001$); mean stenosis diameter from 7.5 ± 3.1 mm to 15 ± 2.8 mm ($p < 0.001$). No major complications occurred.
During the follow up period (mean 4 ± 3 years, range 2 months–9 years), one pt died for complication of aortic valve replacement;.in 23/24 pts, Doppler SPG changed from 61 ± 26 to 25 ± 10 mmHg ($P < 0.001$). MRI/CT scan, performed in 17 pts, confirmed no recurrent stenosis and excluded aortic wall complications. The mean rSBP in the 23 pts dropped from 143 ± 22 to 119.5 ± 14.2 mmHg ($P < 0.001$).
Of 19/23 pts (83%) with arterial hypertension mmHg before the procedure, 16 (70%) have normal 24 hours Holter pressure monitoring values (mean: 116 ± 6 mmHg) at follow-up. Among 15/23 (65%) pts on antihypertensive medications before the procedure, in 12 pts (80%) the therapy was withdrawn 6 ± 9 months (2 days–18 months) after the procedure and 3 (20%) are still on therapy.
Conclusions: In our experience stent dilatation of native and recurrent coarctation is an effective therapy. In the majority of patients the arterial pressure normalizes and aortic wall complications at mild-long term follow-up are rare. An accurate evaluation of the changes of arterial pressure and of aortic wall complication with imaging techniques is mandatory.

P-123
Transcatheter Duct Occlusion in Adult Patients
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Introduction: Treatment of adults with patent ductus arteriosus (PDA) is sometimes complicated by risk factors such as pulmonary hypertension, large ductal dimension or calcification.
Methods: Retrospective study of transcatheter occlusion of PDA in adults.
Results: Among 385 procedures for transcatheter occlusion of PDA, there were 46 adult patients, with mean age of 33 ± 13.3 years (78% female). In 41 patients (89%) percutaneous closure was the first attempt at duct occlusion. Five patients (11%) had residual duct after surgery. One patient had a duct too large and was operated. Six patients underwent a second procedure, for large duct and unavailable device at first attempt (1) or residual flow after previous device (5). Devices used included detachable coil(s) (Cook) – 72%, Amplatzer duct occluder (AGA) – 15% and Rashkind device (Bard) – 13%. In 9 patients more than one device was used.
Twelve patients had pulmonary hypertension (PHT). In seven patients trial occlusion with balloon catheter was performed and the PDA was considered untreatable in three. In the remaining patients with PHT (9/12), duct occlusion was achieved with reduction of PA pressures. Five patients had duct calcification; all underwent successful transcatheter occlusion.
Mean procedure time including trial occlusion was 105 min, with mean fluoroscopy time of 22 min. In the last 5 years these were reduced significantly. Both times were inferior in children who underwent the same procedure (51 and 8.5 min). Overall, 64 devices were implanted in 42/46 patients, with immediate angiographic occlusion in 25/42. At 6 months follow-up, there was total occlusion in 37/42 (88%) and the 5 patients with residual flow underwent repeat catheterization, with occlusion.
Complications included atrial fibrillation (1), device embolization, retrieved at the same procedure (1) and hemolysis (one patient with residual flow, treated with additional coils). Mean follow-up was 2.2 years (3 months to 14 years) with no late complications.
Conclusions: Transcatheter PDA occlusion in adults is safe and efficient, even in some patients with PHT. Patients with PHT should undergo duct trial occlusion, as there is frequently a

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decrease in pulmonary artery pressures. Duct occlusion may then be performed at the same procedure.

P-124
Self-expanding nitinol stents may improve Right-ventricular-to-Pulmonary artery-Conduit obstructions after Norwood-Operation
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Background: Conduit obstructions due to intimal proliferation or kinking may occur within a few weeks after Norwood Operation and lead to reduced pulmonary blood flow, desaturation and growth retardation of the pulmonary vascular bed. Beside surgical conduit-replacement interventional therapy is an alternative, although balloon-based interventions in cyanotic patients carry a higher procedural risk. Self expanding stents can be delivered into obstructed conduits without relevant additional impairment of blood flow.
Objective: To report our preliminary experience with self expanding stents in RV-PA-Conduit-obstructions.
Methods and patients: Three patients with Hypoplastic Left Heart Syndrome and 5 mm Goretex-Conduit who developed conduit-obstructions after Norwood-procedure underwent cardiac catheterization and received one (n = 2) or two (n = 1) self expanding nitinol stents.
Results: Time interval from Norwood-Operation to intervention was 16 to 20 weeks. The saturations in room-air varied between 63 and 69%. Angiography revealed obstructions at the proximal site of the tubular conduit part with a minimal diameter between 2 and 3.3 mm. Pulmonary vessels were considered yet too small for cavo-pulmonary anastomosis in all cases. Therefore a 6×20 mm selfexpanding nitinol stent (precise™, Cordis, Johnson & Johnson) was delivered over a 0.014 floppy-wire through a 6F short-sheath via femoral venous access. One stent "jumped" too far into the distal conduit, so a second device was placed proximally overlapping the first stent. There was no procedural complication, stent expansion was well tolerated. There was an indentation in each stent immediately after implantation, which disappeared completely after 48 hours in all patients. Improvement in arterial oxygenation after 48 hours was seen in all patients and varied between 11 and 17% . In all patients Glenn-operation could be done successfully after 3 to 5 weeks and removing the stented conduit was no problem.
Conclusion: Stenting an RV-PA-conduit with obstruction due to pseudo-intimal proliferation using self expanding nitinol stents may be an alternative treatment to surgical conduit-replacemant with considerable risk and encouraging results and may allow growth of the pulmonary vascular bed to an adaequate size for cavo-pulmonary anastomosis.

P-125
Interventional management of pulmonary sequestration
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Introduction: Pulmonary sequestration is an uncommon entity that may be part of Scimitar's syndrome and can be associated with both intracardiac and extracardiac malformations. The abnormal systemic arterial supply may impose an important volume load on the heart. Embolization of this feeding artery is an alternative for surgical management.
Methods: Retrospective study of all children with pulmonary sequestration that underwent interventional embolization in our institution.
Results: In a 9 yr period 7 pts (6 infants age 1–5 mo, 1 boy age 4 yrs) underwent embolization of the systemic feeding artery of the lung sequester. All 6 infants were in heart failure. The 4 yr old showed left heart volume overload by echo. Sequesters were all right sided, with variable hypoplasia of the right lung. In 6 the sequester artery originated at the level of the coeliac trunk, in one direct from the aorta at the level of the diaphragm. The sequester drained into a right sided scimitar vein in 3 and right lower pulmonary vein in 4 pts. Additional intracardiac abnormalities were present in 4 pts. Embolization was performed with multiple coils in 6 pts, in one infant two 12 mm amplatzer vascular plugs were placed to occlude the vessel. Complete occlusion was obtained in 6/7 pts. In one 2 mo old patient the sequester had to be removed surgically due to extensive necrosis with septic signs. One pt died of respiratory problems due to tracheamalacia with severe lung hypoplasia. Rapid clinical improvement was observed in all other pts. 3 pts underwent cardiac surgery later (scimitar vein reimplant and ASD closure in 2, VSD closure in 1). At follow up (8 mo–9 yrs) the clinical condition of the children is good.
Conclusion: Embolization of the sequester feeding systemic artery is a successful alternative to surgery in most patients.

P-126
Safety and efficacy of different minimally invasive atrial septal defect closure
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Background: Secundum atrial septal defects (ASD) have been usually closed with low mortality by a full sternotomy (FS). The technical ease of ASD closure coupled with a desire for a smaller scar has triggered interest in minimally invasive closure (MIC) to obviate the morbidity associated with sternotomy and the psychological impact of a easily visible scar.
Objective: We present our preliminary experience and a comparison between the MIC and FS approaches.
Methods: From January 2006 and December 2006, 34 pts underwent surgical ASD closure. A MIC was used in 14 and a FS in 20 pts. A right antero-lateral minithoracotomy was used in 6 and an inferior Jministernotomy in 8 pts. The only criterion for a MIC procedure was a body weight greater 20 kg. In female the minithoracotomy was preferred while in male the ministernothomy was chosen.
Mean age was 33+/−12 years in the MIC and 21+/−10 years in the FS group. Mean body weight was 51+/−17 kg and 33+/−16 kg respectively. Female were 11 in the MIC pts (100% among mini-thoracotomy approach) and 13 in the FSA pts.
Results: Patch closure was carried out in 82% of pts in the MIC and 90% of those in the FS group (p:ns). Mortality rate was 0%. The average stay in ICU was 0.8+/−0.2 and 1.0+/−0.3 days respectively (p:ns). The postoperative length of stay was 6.5+/−0.7 days in the MIC and 7.1+/−0.8 days in FS group (p:ns). MIC required longer extracorporeal circulation times than FS (46.5 +/−8.6 min vs 39.7+/−5.9 min, $p < 0.01$)
However, there were no differences regarding the length of cross-clamping times (15.8+/−3.1 min vs. 18.8+/−4.4 min, p:ns).

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MIC pts showed less pain, less respiratory discomfort and better cosmetic results.
Conclusions: MIC is an excellent alternative to FS in ASD closure and has to be considered in all young pts. The psychological impact of a long scar has to be taken into consideration as well as the possibility to reduce the respiratory discomfort and the pain.

P-127

Long term results of Fontan operation: twenty years clinical experience at a single medium volume center

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Objective: To assess predictors of mortality in patients with various different forms of functional single ventricle after Fontan operation in a 20 years time frame.
Methods: Patients who underwent Fontan- type operation between 1985 and 2005 were submitted to a retrospective analysis of medical records. Redo Fontan, and one and a half ventricle repair were excluded. Follow up data were obtained from cardiology follow up controls. Statistical analysis was done with statistical programme Stata 9.1.
Results: 116 patients underwent a Fontan operation (M/F= 70/46). Mean age at operation was 4.5 years (median 2.99, range 0.98–32.85 years). Main cardiac malformations were 132 (tricuspid atresia 35, DORV 25, PAIVS 22, DILV 21, HLHS 15, unbalanced AV septal defect 12, HLHC 1, Ebstein anomaly 1). Initial palliative procedure was performed in 102 patients, consisting in a Blalock-Taussig shunt (56), pulmonary artery banding (21), Norwood operation (18), other procedures (8). In 69 patients (59.5%), Fontan circulation was staged, with a bidirectional cavopulmonary connection (50), a hemifontan (13) or a bilateral bidirectional cavopulmonary connection (6). The Fontan type procedure was completed with atriopulmonary or atrio-infundibular anastomosis in 29, intra-atrial flap in 6, extracardiac conduit (EC) in 31, a lateral tunnel (LT) in 50 pts. Fenestration was performed in 55 patients.
Early mortality (<30 days) was 8.6%, decreasing from 22% (1985–1991) to 2.5% (1992–2005).
At a mean follow up of 7.6 years (range 4 months–20.70 years, 94% completeness), 10 patients had a failed Fontan circulation requiring redo Fontan in 6, heart transplant in 2, take down in 2. In addition, 8 patients (6.8%) died late after Fontan, late mortality decreasing from 11% (1985–1991) to 5% (1992–2005).
Prolonged ventilation (HR 5.5) and atriopulmonary connection (HR 3.02) are correlated with a high risk for global mortality (p-value <0.05). Total cavopulmonary connection (HR 0.26), staged approach (HR 0.37) and fenestration (Odds Ratio 0.10) are correlated (p-value <0.05) with a reduction of the mortality.
Conclusions: Fontan circulation has become a satisfactory long term palliation for functional single ventricle anomalies, in terms of early and midterm outcome.

P-128

Integrating post operative value of cardiac Troponin-I for a better correlation with in-hospital outcomes after congenital heart surgery

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Objective: Cardiac Troponin-I (cTnI) is a well-recognized early postoperative marker for myocardial damage in adults and children after heart surgery. The present study was undertaken to evaluate whether the integrated value (area under the curve (AUC)) of postoperative cTnI is a better mode to predict long-term outcome than post operative cTnI maximum value, after surgery for congenital heart defects (CHD).
Methods: Retrospective cohort study. 279 patients (mean age 4.6 years; range 0–17 years-old, 185 males) with congenital heart defect repair on cardiopulmonary by-pass were retrieved from our database including postoperative cTnI values. Maximal post operative cTnI value, post operative cTnI AUC value at 48h and total post operative cTnI AUC value were calculated and then correlated with duration of intubation, duration of ICU stay and mortality.
Results: The mean duration of mechanical ventilation was 5.1+/−7.2 days and mean duration of ICU stay was 11.0+/−13.3 days, 11 patients (3.9%) died in post operative period. When comparing survivor and deceased groups, there was a significant difference in the mean value for max cTnI (16.7+/−21.8 vs 59.2+/−41.4 mcg/l, $p<0.0001$), 48h AUC cTnI (82.0+/−110.7 vs 268.8+/−497.7 mcg/l, $p<0.0001$) and total AUC cTnI (623.8+/−1216.7 vs 2564+/−2826.0, $p<0.0001$). Analyses for duration of mechanical ventilation and duration of ICU stay by linear regression demonstrated a better correlation for 48h AUC cTnI (ventilation time $r=0.82$, $p<0.0001$ and ICU stay $r=0.74$, $p<0.0001$) then total AUC cTnI (ventilation time $r=0.65$, $p<0.0001$ and ICU stay $r=0.6$, $p<0.0001$) and max cTnI (ventilation time $r=0.64$, $p<0.0001$ and ICU stay $r=0.6$, $p<0.0001$). Conclusion: Cardiac Troponin I is a specific and sensitive marker of myocardial injury after congenital heart surgery and it may predict early in-hospital outcomes. Integration of post operative value of cTnI by calculation of AUC improves prediction of early in-hospital outcomes. It probably takes into account, not only the initial surgical procedure, but probably also incorporates the occurrence of hypoxic-ischemic phenomena in the post-operative period.

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Acute Neurologic Complications Occurred After Cardiac Surgery In Children

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Background: Despite advances in cardiac surgery including deep hypothermic circulatory arrest and corrective operations in early life, surgical approaches may be associated with neurologic complications in the immediate postoperative period as

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well as with subtle cognitive and motor deficits at long-term follow-up.
Aims: (i) To determine the incidence and spectrum of early postoperative neurologic complications; (ii) to identify predictors of risk of neurologic sequelae.
Methods: We reviewed retrospectively 265 children (age range 0–10 years) who underwent cardiac surgery from February 2005 to February 2006. Patients were divided into 2 groups: (i) with aortic arch obstruction; (ii) without aortic arch obstruction. The following variables were considered: (i) age at operation; (ii) preoperative data: gestational age, birth weight, gender; (iii) pre-existing neurologic deficit or known chromosomal abnormality; (iv) use of extracorporeal circulation (ECC); (v) surgery with deep hypothermic circulatory arrest (DHCA) or low-flow cardiopulmonary bypass (CBP). Positive endpoints were clinical or laboratory findings of postoperative cerebral impairment. Time at onset of neurologic sequelae was recorded from day 0 to day 5.
Results: Neurologic sequelae were found in 6 out of 265 patients (2.3%) with a mean age of 15 months; age range at time of surgery: 4–36 months. In all of these patients, surgery was performed in ECC; in 2 of them, DHCA was used. Neurologic sequelae occurred in 4.2% of children with aortic arch obstruction while 2% occurred in those without. Pre-operative data were insignificant. Neurologic complications were: (i) generalized seizures with epileptic activity in 3 children; hemiparesis in 2; paraplegia in 1 child.
Conclusions: All the neurologic complications occurred during ECC, with an increased risk for children aged three years or younger, and in patients with aortic arch obstruction compared to other congenital heart diseases. Vital parameters and laboratory data did not provide additional information as predictors of neurologic risk factors. Although preliminary, our data suggest that neurologic assessment in the pre-operative stage may be of value in patients with aortic arch obstruction and may help in parental counselling about the risk and prognostic factors associated with the cardiac surgery.

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Conversion of Atrio-pulmonary Connection to Total Cavo-pulmonary Connection: A Single Centre Experience with the Fontan Circulation
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Background: Conversion to total cavo-pulmonary connection (TCPC) with rhythm surgery is an accepted surgical strategy in patients with failing Fontan procedures involving an atrio-pulmonary connection. We reviewed our experience with this particularly difficult group of patients.
Methods: A retrospective analysis of baseline characteristics and outcomes of all patients undergoing TCPC in the course of a failing classical Fontan at our tertiary care centre was performed.
Patient and procedural characteristics: 9 patients (5 males, 4 females) underwent TCPC between 2002 and 2006. Indications for Fontan conversion were intractable atrial arrhythmia in 3 patients, hemodynamic compromise in 3 patients, and worsening protein-losing enteropathy (PLE) in 3 patients. The mean age at TCPC was 21 years, and the mean interval after the initial Fontan operation was 16.3 years. The surgical procedure used a Gore-Tex-Graft as an extracardiac conduit and implantation of a pacemaker in all patients, combined with rhythm surgery in 6 patients. In 5 patients a fenestration between the conduit and the atrium was created.
Results: One patient with severe PLE and preoperative functional class NYHA IV died the day after surgery due to severe cerebral edema, probably due to intraoperative air embolism as a consequence of veno-venous collaterals. The average hospital stay of the surviving patients was 36 days, with a mean stay on ICU of 18 days. Mean follow up time was 16 months. One patient without rhythm surgery had recurrence of atrial fibrillation on the 9th postoperative day. NYHA functional class improved in all but one patient by at least one class. In 2 patients the fenestration between the conduit and the atrium had to be closed during follow-up due to persistent low systemic oxygen saturation.
Conclusion: TCPC is an effective treatment of atrial tachyar-rhythmias in patients with a failing Fontan circulation, even in those with PLE. Rhythm surgery should be an integrative part of the procedure. However, patients undergoing this intervention are at high risk to suffer various peri- and postoperative complications. A long ICU- and hospital stay must be anticipated. Despite these limitations, TCPC is feasible with an acceptable perioperative risk in experienced centres.

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Initial experience with unifocalization and repair of pulmonary atresia with VSD and major aortopulmonary collaterals in Lund
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Pulmonary atresia with VSD and major aortopulmonary collateral arteries (MAPCAs) is a complex congenital anomaly whose surgical management has evolved over the years. From no surgery or palliative shunting to complex surgical reconstruction of the pulmonary vasculature with unifocalization. A unifocalization programme was started in 2001. In this study we describe our initial experience with unifocalization.
Method: Hospital records were retrospectively analysed for all patients with PA/VSD/MAPCA referred for surgical evaluation at our institution since 2001.
Material: Since 2001 21 patients has been evaluated for unifocalization. 5 previously known patients were re-evaluated along with 16 new patients. In 16 patients hypoplastic or severely hypoplastic native central pulmonary arteries were found, in 5 they were missing or non-confluent. A total of 69 MAPCAs were evaluated. 4 patients were found inoperable with unifocalization due to irreversible high pulmonary resistance and/or unsuitable anatomy.
Results: 17 patients were accepted for unifocalization. In 2 patients unifocalization was deemed impossible at surgery and only a palliative shunt was performed, in 1 as a desperate attempt to save a severely cyanotic patient due to dissection and occlusion of MAPCA after angiography. The patient died the day after surgery. In 7 patients a staged approach was used with a total of 20 operations. Complete repair with VSD closure was obtained in 6. The VSD had to be fenestrated in 1. In 6 patients one stage unifocalization through a midline sternotomy was used. In 4 the VSD was closed at the same time, in 2 it was left open. 1 patient have later had the VSD closed. 2 patients await surgery. In all 33 MAPCA's were unifocalized, a mean number of 2.5 per patient. Median RV/LV ratio after total correction was 0.6, range 0.41–0.8. In 1 patient balloon dilatation and stent insertion has

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been performed twice because of pulmonary branch-stenosis. 1 patient developed choreoatetosis five weeks postoperatively. Mean follow up is 2 years and 21 days. There has been no mortality in unifocalized patients.
Conclusion: Unifocalization has been performed with low mortality and led to total correction with VSD closure in the majority of the operated patients.

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Extra-anatomic conduits for treatment of adult hypoplasia or interruption of the aortic arch
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Introduction: Aortic arch interruption or severe aortic coarctation with arch hypoplasia in adulthood are rare and represent a surgical challenge, due to massive collateral circulation, eventual adherences from previous surgeries, risk of hemorrhage, risk of ischemic spinal lesion or recurrent nerve injury. The best surgical approach is an open issue. We describe our option for treatment of these lesions.
Methods: We describe 4 patients, two with aortic arch interruption, and two with severe arch hypoplasia associated with recoarctation (1) and native coarctation (1). Their ages ranged from 35 to 66 years, and 3 were male. All had severe systemic hypertension. Patients presented with acute pulmonary edema (2), angina (1), and rupture of cerebral aneurysm (1). One patient had associated coronary disease, aortic valve stenosis and ascending aorta aneurysm.
Surgical approach was through a sternotomy (2), right thoracotomy (1) or left thoracotomy (1). All patients underwent placement of an extra-anatomic conduit of Dacron–Gelseal®, in three cases between the ascending and descending aorta and in one, with a calcified ascending aorta, between the left subclavian artery and the descending aorta. One patient underwent simultaneous aortic valve replacement and ascending aorta replacement with a conduit in supracoronary position and coronary artery bypass graft.
Results: There was no mortality. ICU delay was 34 ± 6 hours and admission length was 10 ± 3 days, with one patient being admitted for 21 days for prolonged fever of unknown origin.
In all patients blood pressure gradient was abolished in the immediate postoperative period. In two patients anti hypertensive medication was suspended and in the remaining two was reduced. With a mean follow up of 2.5 years (6 months to 5 years) all patients are well, with no late complications.
Conclusions: Surgical approach to adult patients with severe disease of the aortic arch and aortic isthmus needs to be individualized. The use of extra-anatomic conduits is safe and efficient and a valid alternative to complex reconstruction of the aortic arch.

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Predictors of Preoperative Neurological Outcome and its Impact on Surgery in Children with Congenital Heart Disease undergoing Cardiopulmonary Bypass
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Objective: To determine predictors of adverse preoperative neurological outcome and the effect of preoperative neurological abnormalities on intraoperative parameters in children with CHD.
Design/Methods: From 2004–2006, 141 prospectively enrolled children aged 0–2 years had a neurological examination before undergoing cardiopulmonary bypass.
Results: A normal preoperative NS was present in 30% of patients, whereas 33% were mildly, 28% moderately, and 9% severely abnormal. Impairment included abnormalities of posture, tone and movement. After adjusting for complexity of cardiac lesions and genetic syndromes, preoperative NS neither differed significantly between bi- and univentricular nor between acyanotic and cyanotic heart defects. Children with genetic syndromes were more likely to have neurological abnormalities ($p < 0.001$). A better NS was associated with a higher weight ($p = 0.02$), length ($p = 0.03$) and head circumference ($p = 0.04$) at birth and a higher head circumference preoperative ($p = 0.004$). Feeding difficulties ($p = 0.004$) and higher preoperative haematocrit ($p = 0.05$) were associated with poorer NS. Within the group of cyanotic CHD, the association between feeding difficulties and neurological abnormalities was stronger than within the acyanotic group. Abnormal NS had no impact on intraoperative parameters.
Conclusions: A high prevalence (70%) of preoperative neurological abnormalities in children with CHD undergoing cardiopulmonary bypass was found. Predictors of neurological outcome were measures of birth and preoperative head circumference. Feeding difficulties and high preoperative haematocrit were associated with abnormal neurological outcome. Adverse NS and cyanotic heart lesion have a potentiating effect on feeding difficulties confirming that feeding is an important marker for neurological integrity and CHD. Preoperative neuronal vulnerability does not affect surgery.

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