56 patients undergoing transcatheter atrial septal defect (ASD) occlusion. The Sideris devices were implanted between 9/93 and 2/96 and the Amplatzer devices between 12/96 and 10/97.

Results:

	Sideris	Amplatzer	p
Number of patients	33	26	
Age (years)	4.3 (1.6 -1.8)	4.6 (2.4 -31)	NS
Weight (Kg)	16 (9.6 -91)	17.5 (10.8 -78)	NS
QP/QS	1.8 (1.5 -3)	1.8 (1.5-3.8)	NS
Echo diameter (mm)	10 (5 -19)	12 (6-25)	NS
Balloon size (mm)	16 (8-23)	15 (7-26)	NS
Fluoroscopy (min)	23 (11-54)	13 (8-49)*	<0.001
Implanted	28	24	NS
Removed surgically	1	1	
Complete occlusion rate at :			
1 month	37%	91%*	<0.001
3 to 6 months	44%	96%*	<0.001

Five Sideris devices could not be implanted and were removed at the time of the procedure, whereas only 2 Amplatzer devices could not be implanted. One device in each series was inadvertently implanted straddling the defect and required surgical removal. The Amplatzer device had embolised to the pulmonary artery, whereas the Sideris device produced cardiac perforation and haemo-pericardium. Both patients recovered completely. Three patients who failed Sideris device implantation were successfully treated with Amplatzer devices. One patient with a Sideris device had a significant residual leak closed with an Amplatzer device.

Conclusions: The Amplatzer device produces higher occlusion rates of ASDs with shorter fluoroscopy times.

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Mechanoelectrical interaction in tetralogy of Fallot. QRS prolongation relates to right ventricular (RV) wall mass and biventricular size

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Aim: To evaluate the correlation between QRS duration and ventricular volumes and wall mass in patients operated for tetralogy of Fallot. Background: Recently a relation between QRS prolongation, predicting malignant ventricular arrhythmias, and right ventricular (RV) size has been documented in patients operated for tetralogy of Fallot. This has not been evaluated with accurate imaging methods.

Methods: Thirty patients with repair of tetralogy of Fallot were studied (mean age 13 ± 3 years, 12 ± 2 years postoperatively). 23 had important pulmonary regurgitation, 3 had important residual pulmonary stenosis (>30 mmHg).Ventricular size was assessed with tomographic magnetic resonance imaging. The amount of pulmonary regurgitation was measured with velocity mapping MRI. QRS duration was derived from a 12-lead ECG obtained within 2 months of the MRI studies. Results: Mean QRS duration was 105 ± 26 ms (range 70 - 150 ms). Right bundle branch block pattern occurred in 28 patients. Atrial or sustained ventricular arrhythmias were not documented. QRS duration did not differ between patients with or without restrictive RV physiology. Mean ventricular volumes were: RV end-diastolic (ED): 113 ± 38 ml/ml^2 body surface area; LVED: 79 ± 16 ml/m^2. Mean RV wall mass was 29 ± 9 gr/m^2, mean % pulmonary regurgitation (PR) 28 ± 19 % of RV stroke volume. QRS duration correlated best with RV mass (linear regression: r=0.71, p<0.001). Correlation with ventricular size and pulmonary regurgitation was: RVED: r=0.56, p<0.01; LVED: r=0.50, p<0.01; PR: r=0.43, p=0.02. Conclusions: In patients operated for tetralogy of Fallot, RV dilation alone does not explain QRS prolongation. QRS duration also relates to LV size, but particularly to RV wall mass. This suggests that myocardial factors should still be taken into account as risk factors for ventricular arrhythmias.

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Catheter approach for left accessory pathway ablation: analysis of the pediatric ablation registry data

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The optimum catheter approach, i.e. retrograde (R); transatrial (T); coronary sinus (CS), for radiofrequency (RF) ablation of left accessory pathways (AP) is debatable. The aim of the study was to analyze multicenter (n=29) data to elucidate advantages and limitations of the approaches.

Among 1391 left AP ablations, T was used 966 times; R 462. CS 105 at some time during the RF procedure. Patients: mean age/wt, 12.3 yr/49.5 kg- underlying heart disease (HD), 7%.

A single approach was used in 1258/1391 with success (S) in 96% T, 92% R, and 88% CS. When 1 or 2 more approaches were used after the first approach failed (F), S was 84%. When using only 1 approach, S correlated (multivariate analysis, p<0.05) with experience (E), lateral AP; failure (F) with HD, posterior septal (PS) AP. When T was tried anytime during the procedure, independent correlates of S/F were; EIHD, PS. Similarly for R, correlates of S/F were: none/PS, and for CS: PS/lateral. For fluoroscopy time (FT), T/CS added mean 10.3/ 9.2 min but R none. Regardless of approach, FT was longer with HD, higher weight, PS, and shorter with E. Major complications (MC) were less with E, greater with younger patients, but unaffected by catheter approach. Conclusion: E is an important factor for high S, short FT and low MC regardless of approach. While S is demonstrated for each approach, greater S can be expected when more than 1 approach is tried. The CS approach should be considered early in the procedure for PS AP.

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Extraction and revision of permanent transvenous pacing leads in children and guch

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Transvenous pacing in children is being increasingly used because of its simplicity, better lead characteristics and improved technology. Because of patient growth, transvenous leads need revision; moreover, non-functioning leads must be removed to avoid tissue damage and haemodynamic problems.

Between 1981 and 1997, 147 patients with a mean age of 8.5 years have been transvenously paced. 14 patients have required lead revisions mainly because of child's growth. The interval between implant and-first revision was a mean of 2.3 years. A total of 47 leads have been extracted, 12 atrial and 35 ventricular. The indications for extraction included exit block in 23, short leads in 5, infection in 11 and miscellaneous indications in 8. The method of extraction included traction, commercial explantation systems as

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well as a locally devised technique for extraction through the femoral vein.
There were no major complications either in the revision or extraction groups. 4 leads were damaged during the revision: 2 were replaced and 2 were repaired. The main complication with extraction has been a residual lead tip either in the heart or in one of the veins.
Transvenous leads need servicing especially in children because of growth. Non-functioning leads must be extracted and we describe our experience, including our own technique of extraction through the femoral vein which is more suitable to children compared to commercial kits.

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Recurrence of congenital heart disease in fetuses with familial risk, screened prenatally by echocardiography

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Objectives of the study: to evaluate the recurrence of congenital heart disease (CHD) in a population of pregnant women with familial risk, screened prenatally.
Material and methods: retrospective study of the data of 591 pregnant women with familial risk for CHD studied in our Center in the period November 1984 - October 1997 by means of fetal echocardiography. Eight pregnant women were followed-up in two consecutive pregnancies. 550 cases had single familial risk, 396 of them had one previous child with CHD, mother alone was affected in 64 cases, father alone in 29 cases, other relatives in 61 cases. In 41 cases there was a multiple risk (2-4 relatives). Fetal cardiological diagnosis was compared with postnatal findings at follow-up.
Results: CHD was found in 29/591 infants, with total recurrence of 4.9%: in 25/550 (4.51%) of cases with single familial risk and in 4/41 (9.7%) of cases with multiple risk. The recurrence of CHD was 4.5% (18/396) when one, I previous child was affected, I 1. 8% (2/17) when 2 previous children had CHD. When mother alone was affected, the recurrence was 3.1% (2/64), while mother and another relative affected had recurrence rate 1/8 (12.5%), with a specific recurrence of ostium secundum atrial septal defect (ASD 11) and of hypertrophic cardiomyopathy (HCMP). When father alone was affected, the recurrence was 2/29 (6.8/o), with specific recurrence of Tetralogy of Fallot (TF) and of ASD 11, while with father and another relative affected, CHD occurred in 1/8 (12.5%). In case of less direct relatives the recurrence was 3/61 (4.9%). Specific recurrence of a similar type of pathology occurred in some index cases with hypoplastic left heart syndrome, tricuspid and pulmonary atresia in situs visecrum inversus, atrioventricular defect, ventricular septal defect, TF, ASD II and HCMP- in this latter pathology global recurrence rate was 7/23 (30%) and specific recurrence 6/23 (26%).
Conclusions: Our data show a higher recurrence rate of CHD than in previously published fetal data and confirm the specific transmission of certain ,CHD in some families to indicate genetic predisposition.

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Arterial wall thickness and mechanics of the carotid artery in Williams syndrome

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As Williams Syndrome patients (WS) have a defect in elastin, a major component of the arterial wall, with frequently associated hypertension, we looked for morphologic and physiologic abnormalities of the arterial wall of the carotid artery in 8 WS using a height resolution vascular ultrasound technique with computerized analysis (Iotec system, Io Data Processing Co, Paris, France). WS group was compared with a control group of same age (10.4 ± 6 vs 10.6 ± 4), weight and body surface area.
WS group had higher systolic (137 ± 14 vs 109 ± 10 mmHg; $p<0.0001$) and diastolic pressure (71 ± 14 vs 60 ± 6.5 mmHg; $p<0.02$). WS had a thicker arterial wall (0.68 ± 0.10 vs 0.50 ± 0.04 mm; $p<0.0001$), and an increased wall thickness/diastolic diameter ratio (0.31 ± 0.09 vs 0.2 ± 0.03; $p<0.0001$). Arterial distension (systolic-diastolic/diastolic diameter) was higher in WS (26 ± 10 vs 19 ± 4%; $p<0.02$). However, cross sectional compliance (1.05 ± 0.4 vs 1.29 ± 0.5 $m^2.kPa^{-1}.10^{-6}$) and distensibility (61.5 ± 22 vs 61 ± 20 $kPa^{-1}.10^{-3}$) were similar in the two groups. Despite the increase in aortic pressure, the wall stress was lower in the WS (32.5 ±10.7 vs 41 ± 8 kPa; $p=0.02$) because of the increase in wall thickness. Therefore the elastic incremental modulus did not differ from the control group (15 ± 99 vs 180 ± 57 kPa. 10^{-3}).
The alteration of the vascular properties cannot account for systemic hypertension in WS as the mechanical function of the arteries are preserved in this condition in spite of the reorganization of the vascular structure.

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Problems identified when screening for familial cardiac diseases in a designated paediatric cardiology genetic clinic

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An increasing number of referrals to screen individuals for familial cardiac disease lead to the creation of a designated joint paediatric cardiology genetic clinic in 1995. An audit of this service has highlighted potential problems for the future. **Resources:** Familial conditions such as hypertrophic cardiomyopathy (1 in 500) and Marfan syndrome (1 in 3-5,000) are relatively common in the general population. In our catchment area of 5.25 million over 1 1,000 people may be affected with just these two conditions. Suspicions of such conditions in a family lead to a request of between 1-5 (median 3) family member being assessed. Referrals for Marfan syndrome screening continue to increase by 200% over 30 months. The relative numbers of patients referred to our clinic still remain a fraction of the potential affected within our catchment area. Waiting times for first clinic appointment are increasing. Additional resources are therefore necessary to meet an increasing demand for screening. **Failure of Proband diagnosis:** Relatives are sometimes referred for screening because a distant family member, who is either dead or geographically remote, is thought to have been potentially affected with a familial cardiac disease. It was not possible to achieve a Proband diagnosis in 19% of all families referred despite requests for death certificates, Coroners reports or medical records. With stricter criteria pro-

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