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Cardiology in the Young

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Use of anaesthesia delivered through a laryngeal mask for transoesophageal echocardiography

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Background: Transoesophageal echocardiography in children and adults with congenital heart disease is usually performed under general anaesthesia using tracheal intubation. Delivery via a laryngeal mask has been introduced as an alternative to the face mask or tracheal intubation for maintenance of the airway.
Objective: To determine the feasibility of conducting transoesophageal echocardiography using a laryngeal
mask airway.
Design: Prospective clinical trial
Setting: Tertiary referral centre.
Patients: Transoesophageal echocardiography was performed in 21 patients while anaesthesia was delivered via a laryngeal mask There were 12 males and 9 females, the age range was 52 months to 69 years (median 10.5 years), and the weight range 16 kg to 78 kg (median 28 kg). An appropriate size laryngeal mask airway was inserted in all patients at the first attempt. Anaesthesia was induced with propofol, and maintained with an inhalational mixture of oxygen, nitrous oxide and isoflurane Patients were allowed to breathe spontaneously during the echocardiographic study. Monitoring involved electrocardiography, noninvasive measurement of blood pressure, pulse oximetry and assessment of inspired oxygen concentration The anaesthesia was maintained for an average time of 24 minutes (range 4 mins to 45 mins).
Results: Transoesophageal echocardiography was successfully performed in 20 patients in whom the hemodynamics remained stable. One patient required intubation after 4 minutes because of cardiovascular instability. Laryngospasm, cough at induction, postoperative airway or pulmonary complications were not seen in any patients. Of the 21 patients, 19 were discharged on the same day, while 2 remained inpatient for subsequent cardiac surgery.
Conclusion: The laryngeal mask provides a safe and secure airway, and could replace tracheal intubation for transoesophageal echocardiography in selected patients who require general anaesthesia

Birth prevalence, spectrum and seasonality of congenital heart disease in Malta: 1990-1994

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Objective: To quantify birth prevalence, spectrum and seasonal variation of congenital heart disease in a closed population.
Methods: Retrospective analysis of all patients diagnosed as having congenital heart disease by 1 year of age between 1990-1994.
Results: There were 232 cases of live-borns with congenital cardiac malformations The female:male ratio was 0.8 The total number of live births over this period was 26,501 The incidence of congenital heart disease was 8 8/1000 live births. The ratio of minor to significant to complex lesions was 7.6.6.9:1 The commonest lesions were ventricular septal defect, pulmonary stenosis and tetralogy of Fallot. The percentages of individual lesions were converted to rate per 10,000 live births and compared to 3 recent epidemiological studies There were higher rates of ventricular septal defect, pulmonary valvar stenosis, tetralogy of Fallot and double outlet right ventricle in Malta ($p<0.0001$, p.0003 [Yates continuity correction] and $p=0.01$ [Fischer's exact test] respectively).
35 of 80 minor ventricular septal defects closed spontaneously within 2 years, with 58% of these being diagnosed in the neonatal period and 93% by 6 months of age
Seasonal variation was analysed with Edwards' method (1961) with correction for background seasonal variation in total live births. Total live births with congenital heart disease (minor, significant and complex) for 1990-1994 did not demonstrate any seasonal variation ($p=0$ 16) When analysed separately, a significant peaking was discovered for June and July ($p=0.015$). This remained significant when groups with significant and complex congenital heart disease were summed ($p=0.03$). Analysis by season strengthened the seasonal peak ($p=0.007$). No seasonal trend was discovered for minor congenital heart disease alone ($p=0.6$).
Conclusions. Minor ventricular septal defects are diagnosed very early in infancy in Malta prior to spontaneous closure and the results reflect the true prevalence of these lesions at birth There is a significantly higher incidence of lesions producing obstruction of the right ventricular outflow tract in Malta. The Maltese gene pool may have an inherent predisposition towards these lesions. There is a seasonal variation in significant and complex congenital heart disease. An environmental factor in the first trimester of pregnancy may be adversely affecting predisposed fetuses conceived in September-October. This trimester coincides with the peak of the coldest weather in Malta.

Formal appraisal of postgraduate trainees

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Methods: After a period of 2-4 weeks in post, a formal appraisal was undertaken. This involved a 20 question (negatively marked) multiple-choice questionnaire with 10 slide questions The doctors' skills in clinical examination were marked according to a standard proforma, and an inpatient clerking and a discharge summary was marked in a standardised fashion The examination process was discussed with the doctors to maximise the teaching value of the process in a non-punitive manner. On an individual basis, the doctors were then interviewed to assess their career intentions, and decide how best to use their time in the Congenital Heart Disease Centre to achieve their objectives. During the last month of the

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post, the examination process was repeated Once the doctors had left the Centre they were invited to express their opinion, in writing, from a place of safety[1]

Results:	Clinical		MCQ		Slides		Admission		Discharge			
Max marks	20		100		50		40		10		Totals	
Test Order	1	2	1	2	1	2	1	2	1	2	1	2
Doctor 1	16	17	82	58	34	46	30	29	8	9	86%	72%
Doctor 2	17	13	55	61	30	45	36	28	10	10	67%	73%
Doctor 3	12	14	65	65	37	44	31	32	7	9	66%	73%
Doctor 4	13	17	27	47	31	27	39	33	7	9	53%	60%
Doctor 5	16	17	33	44	35	31	30	23	9	6	56%	55%
Doctor 6	15	17	56	65	35	43	17	35	7	9	59%	77%
Doctor 7	18	20	41	52	39	33	22	16	8	8	58%	59%

5 doctors were eligible to sit the MRCP (part 1 or 2) examination and of these, 3 had failed in the past. 4 subsequently passed within 6 months of placement in the Congenital Heart Disease Centre

Discussion It was pleasing that the majority improved their clinical examination and technical knowledge scores but disappointing that their admission clerking was less well documented and the discharge summaries remained at the same standard - both teaching and cajoling in these areas were obviously ill-directed or ineffective The assessment failed to identify one doctor whose clinical judgement was poor. The pass rate for MRCP was gratifying but not necessarily attributable to the teaching of the Congenital Heart Disease Centre The most obvious benefit of the assessments was the enthusiasm of the doctors and their appreciation of the Centre's concern for their welfare. In addition, the Centre was able to get to know the doctors' abilities and aspirations earlier in their attachment Whilst these assessments are far from perfect they provide a base, on which we hope to build for the structured reviews of postgraduate trainees required in the era of "Calmanism"

Echocardiographic findings and outcome of aortic stenosis diagnosed prenatally

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Treatment of critical aortic stenosis in the neonatal period is associated with a high mortality The potential of left heart structures to support the systemic circulation determines whether a biventricular repair or Norwood approach is adopted

Aims This study aimed to document, echocardiographically, the left ventricular end-diastolic volume, ejection fraction and size of the aortic root in fetuses with aortic stenosis The relationship of such findings to postnatal outcome was also investigated retrospectively.

Subjects· We included 27 consecutive fetuses with a median gestation of 22 weeks (range 18-35). All were diagnosed between January 1991 and December 1995. Exclusions included any fetus with abnormal cardiac connections, aortic coarctation (n=86), and mitral or aortic atresia with a severely hypoplastic left ventricle (n=130)

Results: The values of left ventricular end-diastolic volume ranged from below the 5th to above the 95th centile at all gestations The z-scores of left ventricular end-diastolic volume at the final prenatal scan ranged from -3 6 to +4.8. Of 8 cases studied sequentially, 6 showed a fall of left ventricular end-diastolic volume across normal centiles. At presentation, the ejection fraction was subnormal in 24 cases (89%). In early gestation, the size of the aortic root was within the normal range for all cases but, beyond 28 weeks gestation, all but one fetus had values below the 50th centile (z-score range -2.5 to +0 5).

Outcome. Following the prenatal diagnosis, 15 sets of parents elected to terminate the pregnancy. Two of the remaining fetuses had attempted prenatal balloon aortic valvoplasty, which was successful in one case Twelve babies were delivered, and ten had postnatal interventions

A biventricular repair was undertaken in 7 (5 survivors) and Norwood approach in 3 (1 survivor) Overall, six of the ten babies who underwent interventions survived. The four fetuses with the highest z-scores for size of the aortic root had successful biventricular repair, as did the fetuses with normal initial ejection fraction. One fetus with left ventricular end-diastolic volume below the 5th centile survived with a biventricular circulation.

Conclusions: Following prenatal detection of aortic stenosis, failure of left ventricular growth is a frequent occurrence. The left ventricular ejection fraction is commonly, but not invariably, subnormal. The outcome of aortic stenosis diagnosed during fetal life is better than previously reported. Fetal echocardiography may contribute to the selection of suitable candidates for biventricular versus Norwood repair

Abnormal blood pressure responses, and reduced aortic compliance, in surgically corrected aortic coarctation

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We investigated 36 patients, mean age 134 months, with surgically repaired aortic coarctation, undertaken at a mean time of 34 months previously, by means of exercise testing, 24 hour ambulatory blood pressure monitoring and an assessment of pressor response to a 1ug/kg intravenous injection of isoprenaline Aortic ratios, ratio of aortic diameter at repair site/diaphragm, and compliance were calculated from magnetic resonance imaging, and peak-to-peak gradients were measured at catheterisation in those with abnormal blood pressure responses 55% had abnormal baseline investigations (loss of diurnal variation on ambulatory blood pressure monitoring, exaggerated pressor responses to isoprenaline, or exercise induced hypertension [systolic rise of >50mmHg]) Of the cohort, 28% had ratios of less than 0.75 as determined by resonance imaging, but there was no significant difference in the ratios of those with or without abnormal blood pressure responses. Aortic compliance was reduced in the group with abnormal blood pressure profiles; mean 14 5uL/mmHg (range 7 8-17.8uL/mmHg) The measured invasive gradient correlated poorly with aortic ratio (r= -0 33) and weakly with post-exercise echo gradient (r=0 52) Of those undergoing catheterisation, 29% had resting gradients across their repair site of greater than30mmHg, this proportion rising to 71% following intravenous isoprenaline.

Conclusions: A high proportion of normotensive patients have abnormal blood pressure responses following surgical repair of coarctation These can occur in the absence of anatomical re-narrowing, or elevated gradients at the site of repair as determined at cardiac catheterisation. Magnetic resonance imaging of aortic morphology is a poor surrogate for invasive measurement of gradients. Persisting changes in aortic compliance in these patients may contribute to the development of abnormal blood pressure responses.

Continuous measurement of Doppler echocardiographic signals in the newborn

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Introduction: Intermittent Doppler echocardiography allows rapid and detailed non-invasive assessment of cardio-pulmonary haemodynamics in sick newborns, infants and children Continuous monitoring of haemodynamics by Doppler study has not been feasible until now, but potentially could give important additional information

Aim:

- To develop a system to record precordial Doppler signals continuously
- To assess the feasibility of the system in healthy terms infants
- To determine the most appropriate site for reliable recording of these signals

Method A 4MHz pulsed wave Doppler crystal was mounted into a steerable device which screws into a commercially available, disposable, adhesive transcutaneous fixation ring (Radiometer, Copenhagen) The signals are then processed by a non-imaging Doppler system (Dopstation, Scimed, Bristol) and displayed in both real time and trend. Initial valuation was undertaken in healthy term infants and our goal was a continuous recording for up to an hour.

Results. Reliable recording was feasible only at the pulmonary trunk Measurement of ascending aortic flow via the suprasternal notch was too difficult due to poor adhesion of the mount. Mitral and tricuspid signals, recorded from the apical area, disappeared periodically due to excursion of the ribcage during respiration. Recordings from the pulmonary trunk were achieved for up to one hour, permitting continuous display of right ventricular output The diameter of the pulmonary trunk was determined from cross-sectional echocardiography Signals were transiently lost during recording with movement, but returned once the baby had settled. In 6 well term infants (birthweight· 2950g - 3620g, age: 14 hrs - 65 hrs) baseline right ventricular output was 172mls/min/kg bodyweight (Range 146-199mls/min/kg) Values were remarkably stable, lying on average within ±16% of baseline value 95% of the time

Conclusion: Continuous recording of Doppler signals from the pulmonary trunk is feasible in healthy term infants Further evaluation of the system is planned during intensive care and pharmacological intervention

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Expression of endothelial nitric oxide synthase in vascular obstructive lesions

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Background: The degree of narrowing of ventricular outflow tract and the great arteries determine the morbidity and mortality of any congenital cardiac malformation. The pathogenesis of vascular obstructive lesions, and the factors responsible for their recurrence, is not well understood. The endothelium as an organ regulating the vascular tone, by synthesizing and metabolizing potent vasoactive substances, like nitric oxide and endothelin, has only recently been recognized. Damage to the endothelium signals proliferation of smooth muscle cells which normally show no proliferative activity.

Hypothesis: Nitric oxide is an endothelially derived vasorelaxant, and inhibits proliferation of smooth muscle cells. Localized absence, or decreased expression, of nitric oxide synthase in the endothelium could therefore lead to proliferation of smooth muscle, resulting in vascular obstructive lesions. Endothelial nitric oxide synthase, the enzyme that regulates nitric oxide synthesis, is a distinct gene produce of 140 KDa, and can be demonstrated semiquantitatively by immunohistochemistry.

Method: We examined the expression of nitric oxide synthase in paraffin embedded sections of aortic and subaortic tissues obtained from patients who have undergone corrective surgery for supravalvar and subvalvar aortic stenosis or aortic coarctation, using endothelial monoclonal antibody to nitric oxide synthase. The presence of endothelium in the sections studied were confirmed by staining for factor VIII related antigen. The staining characteristics for nitric oxide synthase and factor VIII antigen were quantified as '-' or absent, '±' or patchy, '+' or mildly positive, '++' or moderately positive, and '+++' or strongly positive.

Results: All tissues from both patients and controls stained strongly positive (+++) for factor VIII related antigen confirming the presence of endothelium in the tissues studied. Nine patients with aortic coarctation showed mildly positive staining (+). Three patients with subaortic stenosis and two with supravalvar aortic stenosis showed patchy (±) or no staining (-) compared to controls (++). The vessels of the arterial media (vasa vasorum) of all patients showed patchy or no staining (-, ±) when compared to strongly positive controls (+++).

Conclusions: We suggest that a localized defect in the expression of endothelial nitric oxide synthase, causing segmental deficiency of nitric oxide, plays a significant role in the pathogenesis of vascular obstructive lesions and their recurrence.

Competitive pulmonary blood flow prevents the development and progression of intrapulmonary arteriovenous shunting in patients with the superior cavopulmonary anastomosis

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Background. Macroscopic pulmonary arteriovenous malformations are seen in chronic liver disease and following the superior cavopulmonary shunt where the hepatic venous effluent is removed from the pulmonary circulation. A proposed common etiologic factor is the loss of 'hepatic factor' from the pulmonary circulation. In clinical practice, however, only a minority of patients develop evidence of macroscopic pulmonary arteriovenous malformations.

Hypothesis: If the loss of 'hepatic factor' is the sole cause of pulmonary arteriovenous malformations following cavopulmonary shunt, all patients without competitive pulmonary blood flow should show evidence of pulmonary arteriovenous shunting before developing gross pulmonary arteriovenous malformations, and the degree of such shunting should depend on the relative proportion of pulmonary blood flow containing hepatic factor.

Methods We studied 17 patients with cavopulmonary shunt performed at 1-45 months of age (mean 20 5) 15-64 months post-operatively (mean interval 34 months). All patients underwent cardiac catheterisation to exclude venovenous collaterals. They then underwent pulmonary perfusion scan using ^{99}Technetium labelled albumen microspheres to quantify the intrapulmonary right to left shunt. The results were compared to 5 controls.

Results. All patients with a cavopulmonary shunt showed the presence of intrapulmonary shunting. The degree of shunting was significantly higher in the group without competitive pulmonary blood flow (11-64% shunt, mean 32 91%), when compared to the group with additional source of pulmonary blood supply (10-17% shunt, mean 12%) P=0 0076. There was no significant difference between those with additional pulmonary supply and the control group, (3-7% shunt, mean 5 4%). The shunt percentage increased significantly with time only in those without competitive blood flow (shunt % Vs shunt-study interval for the groups of shunted patients P=0 027).

Conclusion: Following cavopulmonary shunt, patients without competitive pulmonary blood flow develop arteriovenous shunting which may progress to pulmonary arteriovenous malformations. This may be due to sustained and inappropriate vasodilation resulting from absent or decreased levels of an inhibitor substance in the superior caval venous blood, when it is the only source of pulmonary blood flow. Augmenting cavopulmonary shunt with an additional source of blood flow containing hepatic factor limits the degree and progression of intrapulmonary arteriovenous shunting.

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