Volume 9 • Supplement 1 • Pages 1-56
Cardiology in the Young

Abstracts of the XXXIII Annual Meeting of The Association for European Paediatric Cardiology, Dublin, 10-13 June 1998

Fetal Cardiology

01

Neurological morbidity after fetal supraventricular tachyarrhythmia

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Background. Pharmacotherapy is used in fetal supraventricular tachyarrhythmia (SVT) to prevent fetal heart failure (HF), hydrops and death. We report on the occurrence of associated cerebral damage in patients (pts) with fetal SVT.

Methods. Fetal SVT was encountered in 19 pts, 8 showed fetal hydrops. All of the 11 pts without hydrops converted to sinusrhythm and survived neurological uncompromised. In the pts with hydrops, treatment was eventually successful in the achievement of sinusrhythm and survival, but in 3 pts cerebral damage was encountered at birth.

Results. Three pts showed signs of cerebral hypoxic-ischemic and/or haemorrhagic lesions on cranial ultrasound in the first hours of life, indicating that pathological events causing these lesions must have taken place in utero. All 3 developed severe FW and hydrops secondary to fetal SVT and lack other causes for cerebral damage. Therapy resulted in incomplete control of heart rate and pts were exposed to rapid changing heart rates, due to frequent relapses into paroxysmal SVT.

Conclusion. We believe that hydropic fetuses with SVT are at risk for development of hypoxic-ischemic or haemorrhagic lesions, due to severe fluctuations in cerebral perfusion. It is of utmost importance to realise this treating fetal SVT and aim at immediate complete control of the tachycardia.

02

Clinical course and outcome of fetal right obstructive disease: a retrospective study of 51 cases

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A retrospective study was undertaken to evaluate the clinical course and outcome of fetal right obstructive heart disease (FROD, n=51) in our Prenatal Cardiology unit since 1988 and analyse our prenatal strategy of management. The fetal population included 32 tretralogy of Fallot (TOF), 13 pulmonary atresia with intact septum (PAIS), 6 Ebstein with pulmonary atresia (EBPA). The TOF group (n=32) was characterized by: age of reference=28, 3 ± 6 wks (range 18 to 39), referral diagnosis was cardiac in 62.5%, other reasons were by frequency order, isolated or combined, extracardiac malformations, IUGR, single umbilical artery, hydramnios, diabetes, all TOF were isolated except 3(AV Canal, dextrocardia, ostium secundum). Genetic and extracardiac anomalies were found in 59%. Genetic anomalies were found in 31 % (T21=5, T18=1, monosomy q18=1, 22 ql 1.2=2, deletion chromosome 6=1). Extra cardiac malformations were found in 28%. Termination of pregnancy (TOP) was performed in 40%(n=13) of TOF for extracardiac reasons, 2 with polyformation died in utero and 1 at birth. The anatomic form was regular in the survival group (n=16,50%), 11 had surgery: 5 had 2 stage surgery, 6 one stage correction (age of palliation =1, 5 to 6 months),(age of correction=6 to 18 months). Survival rate was 82% (delay 2 to 11 years).

The PAIS group (n=13) showed : age of reference 26.5 ± 5.3 weeks (20-36) reference diagnosis was cardiac in 87%. Genetic and extracardiac anomaly were found in 0.2% (T 21 in one). TOP was performed in 9 (4 were unipartite ventricles), 1 died at birth and 3 post surgery (2 valvectomy and 1 shunt).

The EBPA group (n=6) was characterized by : age of reference,= 27 ± 7.6 (17 to 37), in 100% the reason for referral was cardiac, none had genetic anomalies. All had a hypoplastic RV and pulmonary flow ductal dependant. The mortality was 100% (5 postnatal deaths, 1 death in utero). *In conclusion:* In all groups, the referral rate for "cardiac anomaly" was high demonstrating expertise in the primary and secondary levels. The 59% rate of prenatal extracardiac anomalies in TOF is in contrast with a low (0.2%) or absent rate in the other groups. However the prognosis of the surviving TOF is similar to TOF detected postnatally. The necessity of delivery of TOF in specialized centers is questionable in view of the age of palliation. Parental counselling should take into account the ominous prognosis of EBPA and PAIS.

https://doi.org/10.1017/S1047951100011914 Published online by Cambridge University Press

03

Physiology of the fetal right ventricle

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The fetal right and left ventricles (RV, LV) are assumed to function with equal efficiency but the RV functions poorly as a pressure pump postnatally, failing when there is pulmonary hypertension.
Aims: To investigate the normal physiology of the fetal RV.
Methods: Cardiac morphology and function were studied in 20 normally growing fetuses from 20 week's gestation to term using ultrasound and pulsed wave Doppler (PWD). Concurrent estimates of preload and afterload were obtained non-invasively by combining ultrasound, PWD and 2 pairs of electronically phase locked echo-tracking calipers to record pulse diameter wave and volume blood flow in the fetal descending aorta and inferior caval vein.
Results: Ventricular preload increased with increasing gestation and was paralleled by increasing RV & LV force ($t=9.02$ & 8.76; $p<0.001$). However the LV showed a progressive increase in passive filling assessed by velocity time integrals (VTIANTI total) through the mitral valve ($t=-2.75$, $p=0.02$), not seen in RV. Additionally, PWD in the pulmonary artery revealed forward flow in diastole (FFDPA) in 11 indicating restrictive ventricular physiology, but this was never seen in the fetal ascending aorta. No differences in ventricular loading conditions in these fetuses as assessed by pulse wave velocity or volume blood flow were seen.
Conclusions: These observations suggest different physiology of the fetal RV during normal development in comparison to its parallel left ventricle.

04

Early clinical experience with fetal cardiac intervention in human fetuses with severe semilunar valvar obstruction

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The purpose of this study was to review the early experience with fetal cardiac intervention in 14 human fetuses with severe semilunar valvar obstructions. In these fetuses, percutaneous ultrasound-guided fetal balloon valvuloplasties were attempted in order to alleviate their valvar obstruction.
We attempted to collect the following data from video recordings and patient charts: type of semilunar valvar obstruction, gestational age at detection and at intervention, evidence of endocardial fibroelastosis, hydrops fetalis as well as complications of the procedure and fetal demise.
Eight of the 14 fetuses had severe aortic stenosis, 2 had aortic atresia, 2 had pulmonary atresia-intact septum, and 2 had severe aortic stenosis associated with pulmonary atresia-intact septum. In 8 of the 14 fetuses, both gestational ages at initial presentation and at intervention were available. In these fetuses, the mean gestational age at detection was 25.6 weeks (range 20-32 weeks); the mean gestational age at intervention was 29.8 weeks (range 27-33 weeks), and the mean time period between initial presentation and intervention was 4.2 weeks (range 3 days - 9 weeks). At intervention, endocardial fibroelastosis was present in 8 fetuses with aortic stenosis and 2 fetuses with aortic atresia. Hydrops fetalis was observed in 3 fetuses with aortic stenosis and in the 2 fetuses with severe aortic stenosis associated with pulmonary atresia-intact septum. Technically successful balloon valvuloplasties were achieved in 7 fetuses, none of which had an atretic valve. Only one of these fetuses remains alive today. Six fetuses survived the procedure but died from cardiac dysfunction in the first days or weeks after delivery. Two fetuses with pulmonary atresia could not be treated in utero but successfully postnatally. Four fetuses died early within 24 hours after the procedure, one from a bleeding complication, two from persistent bradycardias, and one at postnatal valvotomy following emergency delivery. Sharp-tipped needles caused balloon rupture and loss of catheter parts within the fetal circulation and heart in 7 fetuses.
The early clinical experience of percutaneous ultrasound-guided fetal balloon valvuloplasty in human fetuses with semilunar valvar obstruction has been poor due to worst case selection, bleeding complications, persistent bradycardias as well as technical problems during the procedure. Technical advances in interventional methods and more careful patient selection may improve outcome in future cases.

05

Anatomy of the hypoplastic left heart syndrome in the fetus

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A retrospective study was performed of fetal heart specimens with the Hypoplastic Left Heart Syndrome (HLHS). The study aimed to describe the anatomic spectrum in fetal HLHS and to define features relevant to diagnosis, management and etiology. 128 fetal specimens with either mitral and aortic (MAA), or aortic atresia and patent mitral valve (AA) were examined. These formed 53% of 240 prenatal diagnoses of HLHS made between 1983 and 1995. In comparison to continuing pregnancies with HLHS the pathological series was biased away from twin pregnancies (8.3% vs 0.5%) and towards fetuses with extracardiac (0% vs 2.9%) and karyotypic abnormalities (0% vs 3.4). Fetuses with MAA showed more systemic (22.4%) and pulmonary venous anomalies (13.4%) than did those with AA (2.1% & 3% respectively). Division of the left atrium (7.5% vs 0%), primary hypoplasia of the interatrial communication (41% vs 0%) and arch anomalies (11.8% vs 4.4%) were also more frequent in MAA vs AA. In contrast, fetuses with AA had a significantly larger aortic roots, showed cellular hyperplasia, calcification and endocardial fibroelastosis of the left ventricle and secondary closure of the flap valve (44%). The mode of aortic atresia was muscular obstruction in 85% of MAA and an imperforate aortic valve in 82% of AA. Preductal coarctation was common (87%) to both groups and tricuspid dysplasia rare (2.6%). The findings show anatomic differences between fetal hearts with MAA & AA that could affect post-natal management and give clues to the etiology of HLHS. Distinction between MAA and AA should be possible by fetal echocardiography.

https://doi.org/10.1017/S1047951100011914 Published online by Cambridge University Press

06

Cardiac changings in fetuses with immunhemolytic anemia and their relation to hemoglobin and norepinephrine levels

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Severe fetal cardiac decompensation and intrauterine death are well known complications of immunhemolytic anemia. Chronic heart failure may lead to norepinephine release with concommitant myocardial hypertrophy. The aim of this prospective study was to determine cardiac dimensions and Doppler derived data in fetuses with immunhemolytic anemia, and to relate them to fetal hemoglobin and norepinephine levels.
Methods: Thirty anemic fetuses due to Rhesus incompatibility underwent a total of 76 intrauterine transfusions. Before cordocentesis, a complete fetal echocardiographic examination was performed, and cardiac dimensions and Doppler flow pattern at all valves were measured. The data were compared with gestational age related reference values. Before fetal transfusion, an umbilical blood probe was obtained for measurement of hemoglobin, epinephrine and norepinephrine. Statistical analysis included descriptive statistics, linear regression and correlation analyses, and Wilcoxons tests for comparison of study and reference collective. A $p<0.05$ was chosen as level of significance.
Results: The mean hemoglobin before the first transfusion was 8.6g% (SD 2.7g%) at a mean gestational age of 28.3 weeks (SD 5.0 weeks). Norepinephrine values did not differ to a reference collective, but were higher than epinephrine values.
The most striking echocardiographic finding was myocardial hypertrophy of all ventricular walls in the anemic fetuses. Mean blood flow velocities at the aortic valve (vmean) were significantly increased, and were negatively associated with the fetal hemoglobin, positively with the norepinephrine value.
Conclusions: Fetal myocardial hypertrophy in immunhemolytic anemia may be a result of an increased cardiac workload, which was found by left ventricular Doppler measurements. This isolated increase of left ventricular vmean reflects the restributional mechanisms with preferential perfusion of the brain. Our results suggest that they may be mediated by norepinephrine. Vmean at the aorta could serve as predictor of the degree of fetal anemia.

07

Diagnostic difficulties, associated abnormalities and outcome in 328 cases of atrioventricular septal defect diagnosed prenatally

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Records of 15,481 high-risk pregnancies assessed by fetal echocardiography between 1980 and 1996 were searched for a diagnosis which included atrioventricular septal defect (AVSD). Prenatal diagnosis, definitive diagnosis, including extracardiac and karyotype abnormalities, and outcome, were evaluated in 328 cases where AVSD was diagnosed either pre- or postnatally.
In 318/328 cases the diagnosis was suspected prenatally and this was confirmed in 296 (90%). In the other 22 (7%) cases, congenital heart disease without AVSD was confirmed in all but 2 cases, in whom autopsy performed outside of specialist centres reported a normal heart. In 8, AVSD was a part of differential diagnosis in addition to other cardiac defects. The remaining 12 had no AVSD but had dilated coronary sinus (4), ventricular septal defect (5) or isomerism without AVSD (3). In 10/328 cases an AVSD was diagnosed postnatally but the prenatal diagnosis had been of left atrioventricular valve atresia in 8 cases and right in 2.
Of 306 cases of confirmed AVSD, 102(33%) were associated with isomerism, 47(15%) with other significant extracardiac anomalies and 107(35%) with a confirmed abnormal karyotype.
Up to July of 1993, 122 of 199(61%) pregnancies resulted in termination, and 26(33%) of those continuing were surviving at latest follow-up. Since then, 58 of 107(54%) elected to terminate and 27(55%) of 49 continuing are currently surviving.
Diagnosis of AVSD may cause difficulties in a small percentage of cases, but this is often due to confusion with other abnormalities. There has been a significant improvement in survival rate over the last few years.

08

Maternal 'lived' experience following prenatal diagnosis of a congenital heart defect

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Fetal cardiology is a pioneering specially, with a potential to allow parents time to come to terms with the diagnosis of a congenital heart defect in their baby. There is a lack of research concerning the emotional impact on the mothers who discover, in pregnancy, that their fetus has a major heart defect. The purpose of this qualitative study was to investigate and highlight the significance the mothers attach to this experience. Phenomenology, an inductive descriptive research method was chosen, whereby human experience is illuminated by the individual's perspective. Six mothers, in whom prenatal diagnosis of a congenital heart defect had been made, participated in the study. Five were interviewed using a formal unstructured technique and recorded on tape, and one gave a written account. All the mothers described their subjective experience in detail. Data was analysed according to Colaizzi's procedure. Over 400 significant statements were extracted from the transcripts. These were arranged into common themes, from which 16 theme clusters emerged, falling into 5 categories.
Within the theme clusters the mothers expressed many similar thoughts and feelings. These included anger, injustice and self-blame for having conceived a baby with a heart defect. They expressed both fear and love for the baby, who they felt was giving them signs. None of the mothers wanted the pregnancy to end, and all had developed coping mechanisms. The 'lived' experience of a prenatal diagnosis of congenital heart defect is a complex phenomenon for the mother. More importance needs to be attached to the emotional follow up of these mothers, rather than concentrating on the diagnosis alone.

09

Improved surgical outcome after fetal diagnosis of hypoplastic left heart syndrome

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Prenatal diagnosis of hypoplastic left heart syndrome (HLHS) provides a unique opportunity to plan perinatal management. Yet, the majority of infants with HLHS are diagnosed shortly after birth, requiring resuscitation and transport to a tertiary care center. Studies from large centers report a survival of 70% after the first, and 90% after the second stage surgical palliation. The influ-

https://doi.org/10.1017/S1047951100011914 Published online by Cambridge University Press