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Pulmonary atresia and intact ventricular (PA-IVS): A Swedish multicenter study

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PA-IVS is an uncommon and heterogenous congenital heart defect. The treatment strategy largely depends on right ventricular size and presence of ventriculocoronary connections (VCC). We analyzed the outcome of all children with PA-IVS in Sweden born 19801996. 69 children (31 girls/38 boys) with PA-IVS were born during the study period. Pregnancy and delivery were normal in 54 cases. 11 children were small for gestational age (SGA). In the initial evaluation 2-D echocardiography and angiocardiography was used in 45 cases and echocardiography alone in 24. VCC were diagnosed in 29 patients. Six children (9%) were not offered surgery; one died at catheterization, five were considered inoperable with one surviving and receiving surgical treatment after the neonatal period. 63 children had surgery within two weeks; aortopulmonary shunt (29pts), aortopulmonary shunt and pulmonary valvotomy (PV) (22pts), PV (5 pts), PV and right ventricular outflow (RVOT) patch (3 pts), PV and RVOT-patch and aortopulmonary shunt (4pts). Twelve children (19%) died within 30 days after the initial procedure and an additional five (8%) died late. Additional surgery was performed in 41 children (1-7 procedures, median 2). Surgical mortality (<30days) in this group was 7% (3pts) with one late death. Mortality in SGA babies was 82% (9/1 1). Mortality in patients with VCC was similar whether right venticular decompression (6/12 pts) was performed or not (8/17 pts). At the end of the study 43 children were alive (62%), 24 of whom with biventricular (VCC in 3 pts) and 19 with univentricar repair (VCC in 12 pts). In conclusion, the outcome in children with PA-IVS is grave with a total mortality of 38%. Main risk factors were intrauterine growth retardation (mortality 82%) and the presence of VCC (mortality 48%). The majority of patients with VCC survived with univentricular repair, but VCC did not preclude biventricular repair.

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Assessment of the pulmonary vasculature under different loading conditions prior to Fontan operation

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Current techniques in the haemodynamic evaluation prior to Fontan operation are frequently limited due to low pulmonary blood flow. SVC flow is about 50-70% of total systemic venous return. This potentially makes pre-Fontan evaluation after CP shunt unreliable. Aim: To develop a catheter technique that allows to assess the pulmonary vasculature under different loading conditions. Methods: In a prospective series of 6 pts with a CP shunt (age 3.6 (1.0) yrs) baseline haemodynamics were obtained. One pulmonary artery was temporarily occluded with a Berman catheter and haemodynamics and angiography were repeated under increased flow conditions to the contralateral lung. Results at baseline were compared with the mean of measurements during temporary occlusion of the right and left PAs. Results: No complications were encountered. There was no significant change in systemic pressure, atrial pressure or systemic SO_2. PA diameter during occlusion increased by 7.9(2. 1)% and PA area by 14.4(6.9)% [P<0.01]. SVC pressure rose from 12.7(1.0) to 15.5 (0.9) mmHg [P<0.01]. Transpulmonary gradient increased from 4.4(0.6) to 6.6(0.9) mmHg [P<0.01]. Assuming constant pulmonary blood flow during balloon occlusion, calculated PVR increased from 2.2(0.5 6) to 3.4(0.5) units [P< 0.01]. Conclusion: This new technique allows for a two-point haemodynamic assessment of the pulmonary vasculature prior to Fontan operation. It is felt that this approach is of benefit in patients with low pulmonary blood flow as it mimics postoperative haemodynamics.

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Expanding indications to selective coronary angiography in children

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The evaluation of myocardial ischemia in children remains difficult. Surgical manipulations of the coronary orifices with their potential complications are now increasing. In parallel, precise anatomic assessment is required in acquired coronary disease, since surgical options for their therapy are now available even in young infants.

Since 1993, 369 patients (8 days-17.2 years) underwent 480 selective coronary angiographies in our institution. Catheterization of the coronary artery (CA) was mainly performed through standard arterial approach. After surgical translocation of the coronary ostia, angiography was obtained by a right coronary catheter alone (Judkins, Amplatz). In patients with ventricular septal defect, the coronary orifices were catheterized through anterograde venous approach when possible,

Selective coronary angiography was indicated for (I), preoperative assessment of CA anatomy in Fallot, double outlet right ventricle, truncus, anomalous origin of the left coronary artery (n= 57); (II), postoperative evaluation when the coronary ostia had been transferred or after coronary injury, switch, anomalous origin of the left coronary artery, surgical coronary injury (n=203); (III), congenital CA malformation. coronary fistula n=12, atresia/stenosis of coronary orifice n=2, pulmonary atresia with intact septum n=8, others 14; (IV), acquired CA disease: Kawasaki n=29, atherosclerosis n=l, post-transplant n=154. Transient myocardial ischemia due to gas embolization in a coronary artery occurred in 2 occasions, Femoral artery occlusion occurred in 3 resolving spontaneously in one and in the remaining 2 after fibrinolytic therapy. Selective coronary angiography is a safe and effective procedure in children and infants. It can be performed routinely in expanding indications.

Congenital Heart Disease in Adolescents

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Evaluation of handicap, disability and the need of rehabilitation in children operated upon for congenital heart disease (CHD)

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Survival after surgery for CHD has increased dramatically during the last decades in spite of more complex defects being operated on. Information on the physical, psychological and social ability is sparse. We estimated the need of rehabilitation treatment in 426

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children aged 1-16 years operated on for CHD. The material comprised all children in this age group operated on for CHD in four separate geographical areas (patients with persistent ductus arteriosus were excluded). 90 patients (21%) of whom 48 patients had Down syndrome were already enrolled in a rehabilitation programme. The physical, mental and social status of the remaining 336 patients were evaluated by means of a questionnaire. All patients were offered a clinical examination by a pediatrician (MB) in order to evaluate their need for enrollment in a rehabilitation programme. 289 (86%) questionnaires were answered. The severity of the heart defect did not differ between participants and the drop cuts. In 162 (56%) families the child was considered to be healthy. 127 (44%) children experienced significant problems and 59 (20%) requested further examination and support. The patients problems were solved by the pediatrician in 20 children. The remaining 35 had a need to see the physiotherapist, psychologist, social worker and the pediatric cardiologist. Important issues were low physical work capacity, impaired motor skills, learning disability and poor selfconfidence. Eleven patients were considered to be in definite need of continuous rehabilitation treatment. In conclusion 101/426 (24%) of children operated upon for CHD need rehabilitation on long term basis. Children with Down syndrome constitute the largest single group 48/101 (47%). The need of a rehabilitation programme for 11 (11%) children was not identified earlier stressing the need of a careful follow-up programme for children operated on for congenital heart defects.

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Myocardial perfusion and function of systemic right ventricle in grown-up patients with corrected transposition of the great arteries

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In specific clinical setting while anatomic right ventricle serves as a systemic one deterioration with the time of the systemic pumping chamber is of common concern.
Thus the aim of the study was: 1. To assess systemic right ventricle (SRV) systolic function in the setting of systemic pressure overload; 2. To attempt identification of the features contributing to SRV failure in pts with corrected transposition (cTGA).
Study population consist of 28 pts (14 M., 14F) with cTGA, with the mean age at the study of 23.5±13.6 and mean follow-up 6.8±5.3 y. 19 pts were operated due to additional lesions at mean age 14.3±12.6 y, 9 were unoperated having only trivial or well balanced lesions. There were 2 deaths: one sudden 1 y after correction and second 8 y after surgery due to Eisenmenger syndrome. At the time of the study 19 pts had Ability Index (Ab.In) 1, 7pts had Ab.In 2 or 3. In 15pts, divided in two groups (gr.), radioisotope first pass and SPECT perfusion studies at rest after one dose of Tc-99m. MIBI were performed. Gr.I: 10 pts with Ab.In 1 and mean age of 15.2± 9.0 y and gr.II: 5 pts with Ab.In 2 or 3 and mean age of 29.7±15.8 y. Two groups were comparable in terms of received surgical treatment..
In gr.I, without heart failure, there were normal perfusion scans and mean SRV EF was 38.6±5.3%. In gr.II, with heart failure, there were broad perfusion defects in anterior and/or inferior wall and SRV EF was lowered down to 28.4±5.4 %.
Conclusions: 1. In asymptomatic pts with cTGA and normal systemic right ventricular EF normal myocardial perfusion were found. 2. Extensive perfusion defects correlated with overt heart failure. 3. Further investigations needs to be performed for detection dynamics of systemic RV failure appearance in correlation with perfusion findings and with regard to preoperative and operative factors and other management issues.

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Inhomogenous prolongation of ventricualr repolarization across the myocardium in patients operated on for tetralogy of Fallot. Evidence of electrical myocardial instability?

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Ventricular arrhythmias and sudden death have been reported in pts. with repaired tetralogy of Fallot (TOF). The aim of the study was to examine ventricular repolarization (VR) time indexes, in terms of both absolute measures and dispersion across the myocardium in young pts. operated on for TOF (32pts.; 19M and 13F, age 11. 1±3.4 years; age at surgery 2.4±1.2 years, duration of follow-up 8.5±3.3 years), as these electrocardiographic parameters have shown to be elective in the identification of electrical myocardial instability and hence in the identification of risk for ventricular arrhythmias. Furthermore, it has also been investigated the possible influence on VR of known negative prognostic factors relative to the surgical approach, age at intervention, and presence of, pulmonary obstruction and/or regurgitation. The data of the study group were compared with those of 22 aged-matched asymptomatic control subjects (14M and 8F, age 12±1.5 years). All the patients of the study group revealed complete right bundle branch block and, from the analysis of ventricular depolarization, as expressed by QRS duration, emerged that it resulted significantly longer in total Fallot group than in the Control group (144.8±18.7 vs 94.4±15.1 msec; $p<0.0001$). Particularly, pts. operated through a right ventricular approach, showed higher values of QRS interval than those operated through combined transatrial-transpulmonary approach (160.4±19 vs 141.3±17 msec; $p<0.05$). All the pts. operated on for TOF exhibit, with respect to control subjects, an inhomogeneous prolongation of VR across the myocardium, as showed by the significant increase in the absolute indexes of VR, JTc (332.3±19.8 vs 312.2±22.8 msec; $p<0.01$), QT (392.1±29 vs 353.4±19.9 msec; $p<0.05$) and QTc (477.1±26.5 vs 424.1±25.8 msec; $p<0.05$) with a concomitant prolongation of the indexes of dispersion of VR time, QTCD (65.8±23.2 vs 39±11 msec; $p<0.0001$), JTCD (69.2±24 vs 46.7±18.4 msec; $p<0.0001$), "adjusted" QTCD (19.5±6.8 vs 15.7±4.8 msec; $p<0.05$) and Tp-Te interval (126.6±15.2 vs 93.7±15.9 msec; $p<0.0001$). These findings could, in part, explain the mechanisms that contribute to the development of ventricular arrhythmias in pts with surgically repaired TOF, as this dispersion of refractoriness has been widely accepted as a predisposing factors in the development of ventricular reentry tachyarrhythmias.

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Aortic coarctation in adults - long term follow-up (up to 40 years) in 86 patients

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Study Aim: description of the outcome of 86 patients with aortic coarctation (CoAo) diagnosed after the age of 15 and in care in our hospital since its opening. Methods: retrospective study, questioning personal physicians, patient and/or its family, administra-

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