Topic 10 – Pediatric and congenital heart disease

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Long-Term Outcome of Patients With Ventricular Septal Defect Considered Not to Require Surgical Closure

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Objective: The purpose was to review the long term outcomes of patients with VSD considered not to require surgical closure during childhood.

Background: Although patients with small VSD have generally been considered not to require surgery, more recent data suggest that a significant percentage of these patients develop serious problems during adult life.

Methods: The patient population consisted of 320 consecutive patients with small VSD followed on 11 years, from 1995 until 2006. Spontaneous VSD closure was observed in 66 patients (20.6%). No patients died.

Results: The mean age at the first visit was 6±3.7 years. Spontaneous VSD closure was observed in 66 patients (20.6%). No patients died.

Defect locations were as follows: perimembranous 192 (59%), trabecular 105 (33%), outlet infracristal 5 (2%), inlet 18 (6%).

The small VSD (<3 mm), a trabecular location and an early age of diagnosis had greater chances to the spontaneous closure.

Left ventricular (LV) size by Echocardiography was normal in 292 (91%) patients, enlarged in 26 patients (8%). In our population 8 patients had aortic regurgitation « LAUBRY-PEZZI » syndrome, 7 of them had perimembranous location of VSD and outlet infracristal location in the last case.3 of them had severe aortic regurgitation and 2 of them had surgical closure and valve repair. 2 patients (0.6%) had an episode of endocarditis.

None had systolic LV dysfunction, and pulmonary artery pressure (PAP) was normal in all patients.

Conclusion: Outcome in well-selected patients with a small VSD is good. Surgical closure does not appear to be required during childhood as long as left-to-right shunt is <50% and signs of LV Volume overload are absent.

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Contemporary treatment of isolated vSD in children: amplatzer vs surgical closure

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Introduction: Isolated hemodynamically significant Ventricular septal defects (VSD) were once solely surgical lesions. Since the introduction of percutaneous devices, the management of isolated VSD has evolved. In our center, Amplatzer devices have been implanted since 1998 and for selected isolated perimembranous and muscular VSD since 2002. We therefore reviewed our institutional practice selectively using percutaneous and surgical approaches.

Methods: We retrospectively reviewed clinical, electrocardiographic and echocardiographic data of all isolated percutaneous and surgical VSD closures since 2002. Pre-operative (preop), immediate post-operative (postop) in addition to 1 month, 6 month and annual postop results were studied.

Results: The following table summarizes demographic data: 37 patients who underwent percutaneous closures and 34 who treated surgically, mean follow-up 42 months. Percutaneous group were older (p<0.01) and bigger (p<0.004). The VSD gradient preop was lower in the surgical group (p<0.004), thus patients were in greater clinical heart failure. There was no mortality in either group. At follow-up there were no differences in the incidence of residual VSD (p=0.34). 2 patients with perimembranous VSD and pre-existing aortic valve deformity increased its level to moderate post-Amplatzer implantation versus none in the surgical group (p=0.45). A permanent pacemaker was implanted for complete heart block following post device closure of a perimembranous VSD in one patient and none in the surgical group.

Conclusion: The surgical results are excellent in this sicker patient population, as reported in the literature. Patient selection remains a challenge to avoid post-percutaneous intervention complications such as heart block and aortic insufficiency in the perimembranous VSD patients. Percutaneous closure of isolated VSD, which avoids some morbidity of open heart surgery should however remain part of the therapeutic armamentarium.

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New assessment of subaortic membrane by transthoracic live 3D echocardiography

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Background: Subaortic membrane (SAM) is a fibrous membrane located below the aortic valve. Severe obstruction and/or lesions of valve leaflets require surgical resection of the SAM. Proximity of the SAM to the aortic valve predicts recurrent SAM requiring reoperation. 2D echocardiography (2DE) failed to describe precisely the SAM and its adjacent structures.

Aim: To assess the accuracy of transthoracic real-time 3D echocardiography (3DE) to describe the SAM.

Methods: Twenty-two patients (median age 8 years, 3 to 40 years) with SAM were prospectively enrolled. 2DE and Doppler assessed aortic regurgitation grade, left ventricular to aorta Doppler mean gradient. 3DE was performed using the matrix probe (x1-3 in patients over 7 years old; x7-2 in younger patients). Q-Lab system (Philips) allowed (1) measurements of 3D sub aortic obstruction surface and distance between SAM and aortic leaflets; (2) description of attachment of the SAM to the mitral valve. Ten patients underwent surgical resection of the SAM. Surgical views were compared with 3DE views.

Results: Aortic regurgitation assessed by 2DE and Doppler was present in 14 patients (grade 1 in 10, grade 2 in 4); left ventricular to aorta Doppler mean gradient ranged from 10 to 60 mmHg (mean 25 +/- 10 mmHg). Sub aortic obstruction surface assessed by 3DE ranged from 12 to 65% (mean 22 +/- 15 %); distance between SAM and aortic leaflets ranged from 0.5 to 12 mm (mean 4 +/- 4 mm). Linear relation between Doppler gradient and 3DE obstruction...
Decreased myocardial capillary density in failing functionally univentricular hearts. Yet, further studies with a larger amount of patients are needed to assert the responsibility of the decreased capillary density for the ventricular dysfunction.

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Evaluation of performance and quality of prenatal diagnosis of congenital heart disease in Indre-et-Loire over a period of 15 years

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Objective: To assess the evolution of prenatal diagnosis (PD) of congenital heart diseases (CHD) for the population of Indre-et-Loire, a French department.

Method: Retrospective analysis of the quantitative and qualitative datas of PD between 2000 and 2005 (period 3) and comparison to studies conducted from 1991 to 1994 (period 1) and from 1995 to 1999 (period 2). The CHD considered as detectable are the major CHD with neonatal symptoms.

Results: Period 3: 65.7% of detectable CHD were screened. Using the criteria of PD of period 1, the percentage of CHD detected for periods 1, 2 and 3 is respectively 43.2%, 66.7% and 79.3%. For transposition of the great arteries: 25%, 62.3% and 60%. For hypoplastic left ventricle: 80%, 87.5% and 94.1%; Tetralogy of Fallot: 10%, 42.8% and 75%; Pulmonary atresia with ventricular septal defect: not studied, 50% and 100%. Severe coarctation of the aorta: not studied, 25%, 40%. Atrio-ventricular septal defect: 77%, 77% and 80%. Diagnoses made by a Pediatric-cardiologist are correct and complete in 80% of cases.

Discussion: Performance of PD of CHD is comparable to those reported in the literature. The improvement of PD over time is due to technological progress, greater experience of ultrasonographers and to the systematic visualization of outflow tract in addition to the four chamber view.

Conclusion: Despite the improvement of PD, some life threatening CHD are not screened before birth and require a specific training program for the ultrasonographers.

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Home INR monitoring of oral anticoagulation therapy in children using a point-of-care INR monitor after an education program

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Introduction: The literature demonstrated that children receiving anticoagulation therapy require more frequent monitoring than their adult counterparts due, in part, to the complexity of their underlying medical conditions. Since June 2008, point-of-care International Normalized Ratio (POC INR) monitor are refund by Social Security in France for paediatric patients. The aim of this study is to evaluate the outcomes of home INR monitoring in children through a standardized education and training program.

Materials and methods: The POC INR monitor is refund for under eighteen patients with a cardiologic or non cardiologic indications. Participating parents had to complete a one-day intensive education and training program.