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Influence of valvular asymmetry and orifice area on aortic stenosis in congenital bicuspid aortic valve: a pediatric three-dimensional echocardiographic study

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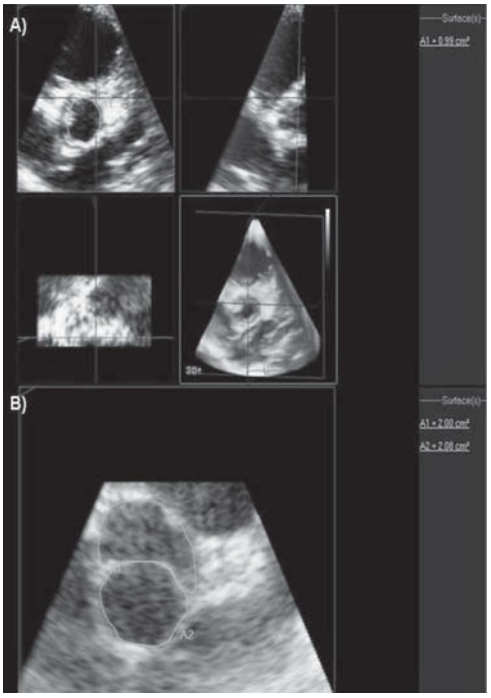
Background: Three-dimensional echocardiography (3DE) was proved efficient to accurately depict bicuspid aortic valve (BAV) which is defined by the presence of two functional cusps because either the valve is really bifoliate or because two cusps are merged by a raphe. Whereas the relationship between the severity of the stenosis and the surface of the aortic orifice was clearly demonstrated in adults, such correlation has never been reported in children.

Objective: The aim of our study was to evaluate, using 3DE, the influence of both valve asymmetry and valve orifice area on the degree of stenosis in case of congenital BAV.

Methods: 70 consecutive children who had BAV and who never underwent surgery or interventional catheterization of the aortic valve, were included in this prospective monocentric study. Using the multiplanar mode review, aortic orifice area was measured in systole (figure, panel A) and aortic cusps surfaces were measured in diastole (figure, panel B) for each patient. The degree of stenosis, estimated by usual echocardiographic methods, was also reported. Results are expressed as the median and first and third quartiles.

Results: Median age was 5.6 years (2.2-11.5). Among the 70 included children, 25 had aortic stenosis. The ratio small/large cusp was strongly associated with aortic stenosis ($p<0.001$). The area under the ROC curve was 0.89 (95% CI, 0.82-0.97). The best cut-off value to differentiate stenotic from non-stenotic valve was 0.75 with 84% sensitivity and 83% specificity. When indexed for body surface area, the aortic orifice area was significantly smaller ($p=0.031$) in case of stenotic BAV (1.51 cm² [0.99-2.28]) compared to non-stenotic BAV (1.99 cm² [1.57-2.52]). Intra- and inter-observer concordances were excellent both for the measurement of the cusps area and for the measurement of the orifice area.

Conclusion: This study allows a better understanding of the functional status of BAV. Aortic stenosis strongly depends on the asymmetry of the valve.



Multiplanar review mode of a bicuspid aortic valve

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Implantable cardiac defibrillator in tetralogy of Fallot

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Background: Implantable cardioverter-defibrillators (ICDs) are increasingly utilized in the growing and aging population with congenital heart disease. Tetralogy of Fallot (T4F) is the most common form of congenital heart disease in ICD recipients.

Methods: We conducted a multicenter retrospective study in 7 centers in France to determine indications and outcomes with respect to death, ICD therapy delivery, and complications in T4F patients implanted between January 2005 and December 2010. A total of 36 patients (mean age 40.7 years; 69% male) were analyzed after a median follow-up of 3 years.

Results: ICDs were implanted for primary prevention in 8 patients (22%) and for secondary prevention in 28 (78%), defined by clinical sustained ventricular tachyarrhythmia, resuscitated sudden death or syncope with inducible ventricular tachycardia. Indications for primary prevention were most often a combination of criteria including right ventricular systolic dysfunction and/or important pulmonary regurgitation (66%), prior palliative shunt (50%), inducible ventricular tachycardia (VT) (39%), QRS duration ≥180ms (25%), non sustained VT (25%). Although 50% of patients implanted in secondary prevention received at least 1 appropriate and effective ICD therapy, only 12.5% received such therapies in the setting of primary prevention (P=0.06). Two patients received heart transplant. Six patients (6%) died during the course of follow-up. Major ICD-related complications occurred in 12 patients (33%).

Conclusion: Patients with tetralogy of Fallot and ICDs for secondary prevention experience high rates of appropriate ICD therapies; however, major ICD-related complications remain frequent. Selection of candidates for primary prevention implantation remains challenging.

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Pediatric cardiovascular disease in Djibouti (Horn of Africa): management and outcome

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Purpose: Few data are available about cardiovascular disease (CVD) in children in the Horn of Africa. We aimed to document the current patterns in Djibouti.

Methods: Clinical features, management and outcome of Djiboutian children between 3 month-old and 15 year-old with CVD were prospectively recorded over a two-year period in our centre (Bouffard French Military Hospital, Djibouti, January 2009 – December 2010).

Results: Clinical examination and echocardiography were performed in 156 patients and 32 (20 %) had CVD. The median age was 5 years; 53% were males. Congenital heart disease (CHD) was observed in 27 (84 %) patients and dilated cardiomyopathy with severe mitral regurgitation in 5 (16%) patients. Ventricular septal defect was the most common abnormality (28 %). Other abnormalities were diagnosed: atrial septal defect (13%), Tetralogy of Fallot (9%), pulmonary stenosis (6%). Three patients had Down's Syndrome and 3 others had multiple congenital anomalies condition. Surgical management was required in 22 (69%) patients and was performed on 15 (47%) cases. Twelve patients were sent abroad for cardiac surgery with humanitarian programs. During follow up (mean 11.3±6.8 months), 5 (16%) patients died. Absence of surgery was associated with significant mortality ($p<0.05$). Age and sex were not associated with mortality.

Conclusions: CVD is at least as common in this Djiboutian community as in other African cohorts of children. The absence of surgery was a major mortality risk factor. Dilated cardiomyopathy was frequent in this study. Much work remains to be done to discover the size and nature of genetic and environmental contributions to these various forms of pediatric heart diseases in the Horn of Africa.

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Population-based evaluation of a suggested anatomic and clinical classification of congenital heart defects based on the International Paediatric and Congenital Cardiac Code

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Background: Classification of the overall spectrum of congenital heart defects (CHD) has always been challenging, because of the diversity of the cardiac phenotypes and the often complex associations. The purpose of the study was to establish a comprehensive and easy-to-use classification of CHD for clinical and epidemiological studies based on the long list of the International Paediatric and Congenital Cardiac Code (IPCCC).

Methods: We coded each individual malformation using six-digit codes of the long list of IPCCC. We then regrouped all lesions into 10 categories and 22 subcategories according to a multi-dimensional approach encompassing anatomic, diagnostic and therapeutic criteria. This anatomic and clinical classification of congenital heart disease (ACC-CHD) was then applied to data acquired from a population-based study of CHD in France, including 2867 cases (82% live births, 1.8% stillbirths and 16.2% pregnancy terminations).

Results: The majority of cases (79.7%) could be identified with a single IPCCC code. The category "Isomerism and visceral heterotaxy" was the only one that typically required more than one code for identification of cases. The two largest categories were "ventricular septal defects" (52%) and "anomalies of the outflow tract and arterial valves" (20% of cases).

Conclusion: Our proposed classification is not new, but rather a regrouping of the known spectrum of CHD into a manageable number of categories based on anatomic and clinical criteria. The classification is designed to use the code numbers of the long list of IPCCC but can accommodate ICD-10 codes. Its exhaustiveness, simplicity, and anatomic basis make it useful for clinical and epidemiologic studies, including those aimed at assessment of risk factors and outcomes. The proposed classification can also provide a structure for various clinical and epidemiologic databases.

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Parental electrocardiographic screening identifies a high degree of inheritance for congenital and childhood non-immune isolated atrioventricular block

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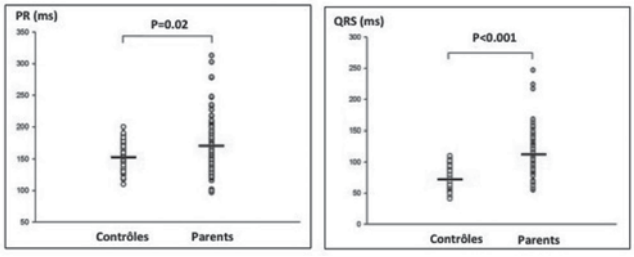
Introduction: The etiology of congenital or childhood non-immune, isolated AV block remains unknown. We hypothesized that this conduction abnormality in the young may be a heritable disease.

Method: A multicenter retrospective study (13 French referral centers, from 1980 to 2009) allowed inclusion of 141 children with AV block diagnosed *in utero*, at birth or before 15 years of age, without structural heart abnormalities and without maternal antibodies. Parents and matched controls were investigated for family history and for ECG screening.

Results: In parents, family history of sudden death or of progressive cardiac conduction defect was found in 1.4% and 11.1% respectively. Screening ECGs from 130 parents (mean age 42.0 ± 6.8 years, 57 couples) were compared to 130 matched healthy controls. All parents were asymptomatic and in sinus rhythm, except one with unknown complete AV block. Conduction abnormalities were more frequent in parents than in controls, respectively found in 50.8% versus 4.6% ($p<0.001$). Long PR interval was found in 18.5% parents but never in controls ($p<0.001$). Complete or incomplete right bundle branch block was observed in 39.2% parents and 1.5% controls ($p<0.001$). Complete or incomplete left bundle branch block was found in 15.4% parents and 3.1% controls ($p<0.001$). Heritability estimate for isolated conduction disturbances was very high, calculated at 91% (standard error=1.019, $p=2.10^{-16}$).

Conclusion: ECG screening in asymptomatic parents from children affected by idiopathic AV block revealed a high prevalence of conduction abnormalities with prolongation of intra-atrial, AV and/or intra-ventricular conduction delay. Heritability estimate confirmed a high contribution of genetic factors. These results support the hypothesis of an inheritable trait in congenital and childhood non-immune, isolated AV blocks.



Comparison of PR interval and QRS complex duration

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Assessment of systo-diastolic ventricular function using tissue Doppler imaging after successful repair of aortic coarctation

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Purpose: the aim of the study was to assess the ventricular systolic and diastolic function after the successful repair of aortic coarctation using tissue Doppler imaging (TDI).

Methods: The study group consisted of 28 patients (mean age 12±4.2 years) after the aortic coarctation (AoC) repair. The TDI parameters and the conventional echocardiographic indices of the left and right ventricular systo-diastolic function were analyzed and compared with the results obtained from 22 healthy controls.

Results: Patients with repaired aortic coarctation had significantly decreased systolic (Sa) tissue Doppler velocities at the lateral mitral (11.8 vs 14.7 cm/s, p=0.001), tricuspid (16 vs 18.7 cm/s, p=0.009), and septal (10.2 vs 12 cm/s, p=0.058) annuli compared with controls. The early and diastolic