Appendix A

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29 Grown-up congenital heart disease: Continuum of care between pediatric and adults cardiologists in Reims University Hospital
A. Charbonneau 1, 3, * , A. Akhavi 1, J.P. Chabert 2, F. Lesaffre 2, D. Metz 2, P. Morville 1, P. Mauran 1, 3
1 Department of pediatrics, American Memorial Hospital, CHU de Reims, France
2 Department of cardiology, Hospital Robert Debré, CHU de Reims, France
3 Department of physiology, Université Reims Champagne-Ardenne, France
* Corresponding author.
E-mail address: acharbonneau@chu-reims.fr (A. Charbonneau)

Background Congenital heart disease (CHD) are seldom cured and the increasing grown-up congenital heart disease (GUCH) population still needs specialized follow-up that has to be organized. In Reims University Hospital, it’s about 10 years ago that pediatric and adult cardiologists initiated a multidisciplinary clinic for follow-up of GUCH patients.

Objectives The aim of the present study is to describe the GUCH population followed in our hospital (CHD, treatments, outcomes, complications, social issues) and the organization of their medical care.

Methods This is a retrospective and observational study including patients with CHD which were over 18 years old in September 2013 and were seen in Reims University Hospital at least once between January 2008 and September 2013.

Results Our GUCH population was 475 patients. Median age was 25.1 years old. Sex ratio was 0.82. Among CHD, univentricular hearts were 10%. Thirty-one percent of patients were taking cardiologic medications. Cardiac surgery had been performed in 62.1%. Interventional catheterization had been undertaken in 19.6%. Regarding the outcome, main complications were arrhythmias (17%), heart failure (5%), pulmonary hypertension (4%), endocarditis (1.2%), death (1.9%). Social supervision was needed in 14.3%. Twenty-six percent of patients had a life partner and 12.4% had children. During the period study, 11.7% of patients were lost to follow-up.

Conclusions After reaching adulthood, patients with CHD need continuous follow-up because complications in GUCH patients are not uncommon. Although transition between adolescence and adulthood is supposed to be at high risk of breaking this follow-up, the present study shows that it is possible to minimize this event. We feel that an organized continuum of care between pediatric and adults cardiologists, as we settled in our hospital, could be an effective way to meet the special needs of GUCH patients.

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30 Infective endocarditis in patients with ventricular septal defect
M. Veyrier 1, A. Soufi 1, M. Bakloul 1, J. Ninet 2, F. Sassolas 1, A. Bozio 3, C. Ducreux 1, S. Di Filippo 1
1 Pediatric Cardiology, Cardiovascular Hospital Louis-Pradel, Lyon, France
2 Cardiac Surgery, Cardiovascular Hospital Louis-Pradel, Lyon, France
3 Corresponding author.
E-mail address: sylvie.di-filippo@chu-lyon.fr (S. Di Filippo)

Summary In the context of recent change in guidelines for prophylaxis of infective endocarditis (IE), the objective of this study was to assess the features and outcomes of IE in children and adults with non-significant ventricular septal defect (VSD).

Methods A retrospective analysis of records of patients with non-significant VSD. Clinical, echocardiographic and microbiological data, and outcomes were assessed.

Results From 1980 to 2013, 57 IE occurred (1 to 4 per year), in patients aged 14.2 ± 11.3 years (med. 12.1), 29 males (51%) and 39 were < 18 years of age. VSD was membranous in all cases, isolated (39 = 68.5%) or associated with mild aortic regurgitation or pulmonary stenosis. VSD was native in 39 (68.5%) and not diagnosed before IE occurred in 4 cases (7%). The cause of infection was unknown in 36% of the cases, while 23% were from dental, 13% from cutaneous, 9% from ENT or digestive origin, and 19% occurred in the early postoperative course of patch closure, i.e. 81% of the cases occurred in native mild VSD. Streptococcus from dental origin was the most frequent causal agent (54.5%), staphylococcus was found in 35% of cases, Gram-negative bacillus in 3.5%. Hemocultures were negative in 7% of the cases. Vegetation was the most frequent echographic lesion, and located either on VSD, and/or tricuspid valve and/or RV free wall and/or pulmonary valve. Aortic valve location occurred in 8 cases. Embolic event occurred in 28 cases (49%): multiple pulmonary embolism in 21 (37%), systemic embolism in 6. Eighteen patients were operated (31.6%): early surgery in 11 (19.3%), delayed patch closure in 7. Six patients died (10.5%). Death was not related to early surgery. FU was 13.4 ± 11.2 years (med. 10.2 years).

Conclusion Infective endocarditis impairs prognosis of mild membranous VSD and dental events are the most frequent origin of infection. Preventive surveillance and management of any dental lesion are probably to be emphasized in these patients.

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31 3D transthoracic echocardiography to assess ventricular septal defect anatomy and severity
K. Hadeed, S. Hascoet *, R. Amadieu, Y. Dulac, P. Acar
Cardiologie pédiatrique, Hôpital des enfants, CHU de Toulouse, France
* Corresponding author.
E-mail address: hascoets@gmail.com (S. Hascoet)

The pathophysiology of ventricular septal defect (VSD) is determined by the size of the defect and the state of the pulmonary
vascular resistance. We assessed the morphology of VSD using 3D transthoracic echocardiography (3D-TTE) and the ability to estimate the pathophysiology.

Methods Forty-eight children with isolated unique muscular (77%) and membranous VSD (23%) were included. Severity of the VSD was rated according to their pathophysiology. (Type 1: minor left-to-right (L-R) shunt; 2a: significant L-R shunt (left ventricular end diastolic diameter (LVEDD) Z-score > 2); 2b: VSD associated with pulmonary hypertension.) 3D VSD measurements were obtained after a multi-planar reconstruction of a TTE 3D full volume (X5-1 or X7-2 matrix probes, iE33, Philips). Diagnosis properties of the ratio of 3D VSD area to aortic annulus area (3DA/AAA), 3D VSD area to body surface area (3DA/BSA) and 2Dmax diameter to BSA (2DD/BSA) were compared.

Results Median age was 8.5 month-old (min 1 max 123). LVEDD Z-score was > 2 in 19 children (40%), (12 (25%) had pulmonary hypertension). 3D VSD systolic area, 3D and 2D max. diameters were correlated with LVEDD. (r = 0.71, r = 0.52, r = 0.55, P < 0.05). Systolic-diastolic variation of 3D VSD area was higher in muscular than in membranous VSD (Median 54% vs 27%, P = 0.0001). VSD were asymmetric with a mean ratio of maximal to minimal 3D diameters of 2.1 ± 1.3 in membranous VSD and 3.2 ± 1.5 in muscular VSD (P = 0.01). VSD severity was correlated with LVEDD Z-score, 3DA/AAA and 3DA/BSA ratio (r = 0.52, r = 0.63, r = 0.60, P < 0.05), but not with the 2DD/BSA ratio. Ability to diagnose type 2b VSD was higher with the 3DA/AAA or 3DA/BSA ratio than the 2DD/BSA ratio (ROC area 0.97 ± 0.01 vs 0.85 ± 0.03). A 3DA/AAA ratio > 0.39 has a sensitivity of 92% and a specificity of 97% to diagnose a 2b-VSD.

Conclusion 3D-TTE allows a morphological and a quantitative assessment of muscular and membranous VSD. The 3DA/AAA ratio is an accurate diagnostic tool to assess the pathophysiology of the VSD.

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32 Right ventricular systolic strain evolution during peri-operative management of congenital heart diseases

Service de cardiologie pédiatrique et congénitale, Hôpital des enfants, CHU de Toulouse, France
* Corresponding author.
E-mail address: hascoets@gmail.com (S. Hascoet)

Background RV systolic strain evolution during peri-operative management of congenital heart diseases (CHD) is unknown.

Methods In this prospective study, RV peak systolic strain (PSS) was measured using 2D speckle tracking echocardiography (Qlab10.0 software, Philips) in 39 children undergoing surgery of a CHD (Median age: 17 months, min. 6 days old, max. 14.3 years old). Three measures were performed the day before surgery, few hours after the surgery and before discharge and compared to conventional echocardiographic parameters of RV and left ventricular (LV) function. The relationships between the evolution of RV-PSS, peri-operative parameters and the type of CHD were assessed.

Results Mean RV-PSS at baseline was −19.5 ± 4.8. RV-PSS was moderately correlated with the heart rate (r = 0.49), the LV Tmad (r = —0.48), the TAPSE (r = —0.54) and the tricuspid S' wave (r = —0.44) (all P < 0.05). RV-PSS was decreased in cyanotic CHD (P < 0.05), in children with congestive symptoms (P < 0.01) and increased in ASD (P = 0.02). RV-PSS was higher in RV volume increased condition such as ASD than in RV pressure increased condition such as Fallot tetralogy (P = 0.006). RV-PSS decreased after surgery (P < 0.0001). Mean difference between pre- and post-operative RV-PSS was 7.5 ± 4.4. The difference was correlated with initial RV-PSS (r = —0.80), the weight (r = 0.54), the ultrafiltration rate (r = 0.43) (all P < 0.05) but not with the duration of aortic clamp, the duration of extracorporeal circulation, the troponin peak level nor the lactates peak level. A higher difference was associated with a shorter duration of mechanical ventilation (P = 0.04) and a shorter stay in intensive care unit (P = 0.03). RV-PSS was better at discharge (median 6 days, P = 0.009) but remained lesser than at the initial exam (P < 0.0001).

Conclusion RV-PSS decrease after surgery of CHD. This decrease seems mainly related to loading condition rather than to RV contractility given its relationship with a faster post-operative evolution.

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