age at repair ($P = 0.02$), a higher systolic pulmonary artery pressure ($P = 0.04$) and a higher grade of LAVVR ($P = 0.03$).

Conclusion.--- PAVSD repair is indicated in adult patients because it carries an excellent prognosis and operated patients have better long-term clinical and hemodynamic status than unoperated patients. Moreover, SWA incidence decreases when surgery is performed before 40 years of age.

http://dx.doi.org/10.1016/j.acvd.2013.06.023

18 Atrioventricular discordance with ventriculo-arterial concordance: Diagnostic challenge, surgical management and long-term outcome

Daniela Laux$^a$, Lucile Houyel$^b$, Fanny Bajolle$^a$, Younes Boudjemline$^a$, Damien Bonnet$^a$

$^a$ Centre de Référence Malformations Cardiaques Congénitales Complexes (M3C)-Necker, Hôpital Necker-Enfants—Maladies, Assistance Publique des Hôpitaux de Paris, Université Paris Descartes, Sorbonne Paris Cité, Paris, France

$^b$ Centre de Référence Malformations Cardiaques Congénitales Complexes (M3C), Department of Pediatric Cardiology, Centre Chirurgical Marie-Lannelongue, 133, avenue Résistance, 92350 Le-Plessis-Robinson, France

Background.--- Atrioventricular discordance with ventriculo-arterial concordance is a rare cardiac defect whose pathophysiology resembles transposition of the great arteries.

Objective.--- To report a series of ten patients with atrioventricular discordance with ventriculo-arterial concordance focusing on segmental analysis, diagnostic difficulties, surgical management and follow-up.

Methods.--- Retrospective review of medical files of all patients with this diagnosis seen from 1983 to 2013 in a single institution. Results.--- Seven patients had (1, D, S) segmental arrangement, two had anatomically corrected malposition (S, L, D) and one had (S, L, S) arrangement. Only five patients were correctly diagnosed preoperatively while diagnosis had to be completed during or after surgery in the other five. Mean age at repair surgery or at last surgical intervention was 3.4 years (range: 5 months–12.8 years). Repair surgery finally performed was atrial switch procedure of Senning or Mustard type in eight of 10 patients. Repair included ventricular septal defect closure in three cases and right ventricle to pulmonary artery conduit or patch in four cases. Main postoperative complications were two cases of baffle obstruction requiring reintervention and one sick sinus syndrome needing pacemaker implantation. There was no postoperative heart block. There were two early postoperative deaths and eight late survivors. Mean follow-up after repair was 6.8 years (range: 5 months–25.4 years) with good functional status in all but one patient who will be listed for heart transplantation.

Discussion.--- Preoperative diagnosis of atrioventricular discordance with ventriculo-arterial concordance remains challenging. Atrial switch procedure is the surgical method of choice. If correctly diagnosed, long-term follow-up is encouraging. Rhythm disturbances and baffle obstruction are the main postoperative problems in this series.

http://dx.doi.org/10.1016/j.acvd.2013.06.024

19 Anomalous left coronary artery from the pulmonary artery associated with other cardiac defects: A difficult joint diagnosis

Daniela Laux$^a$, Claire Bertail$^b$, Fanny Bajolle$^a$, Lucile Houyel$^b$, Younes Boudjemline$^a$, Damien Bonnet$^a$

$^a$ Centre de Référence Malformations Cardiaques Congénitales Complexes (M3C)-Necker, Hôpital Necker-Enfants—Maladies, Assistance Publique des Hôpitaux de Paris, Université Paris Descartes, Sorbonne Paris Cité, Paris, France

$^b$ Centre de Référence Malformations Cardiaques Congénitales Complexes (M3C), Department of Pediatric Cardiology, Centre Chirurgical Marie-Lannelongue, 133, avenue Résistance, 92350 Le-Plessis-Robinson, France

Background.--- Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a coronary abnormality which can be associated to other congenital heart defects which complicates the positive diagnosis of the coronary abnormality, especially before surgery.

Objective.--- Here, we report a series of 13 patients with ALCAPA with a focus on the type of associated heart defect, the moment of diagnosis of the coronary abnormality related to surgery and their outcome.

Methods.--- Retrospective assessment of medical files of all patients with ALCAPA and other congenital heart defects in two important French Departments of Pediatric Cardiac Surgery from 1987 to 2012. Results.--- The heart defect most frequently encountered in association to ALCAPA was aortic coarctation ($n = 5$) followed by tetralogy of Fallot with or without pulmonary atresia ($n = 3$). There was one case of hypoplastic left heart syndrome, one aortic atresia, one right aortic arch, one congenital mitral malformation and one infant with divided left atrium and partial anomalous pulmonary venous return of the right lung. Only four patients had a complete diagnosis of the cardiac defect and the coronary abnormality before surgery. In three cases the coronary anomaly was discovered during surgery conducted for another cardiac defect and treated at the same time by coronary reimplantation. The six remaining patients were diagnosed after cardiac repair. Three of these patients only had a post-mortem diagnosis. Eight of 13 patients died after surgery. Three of them deceased within the first 30 days after repair. The remaining patients are in good health with a median follow-up of 5.3 years (range: 2.1–8.5 years).

Discussion.--- This series confirms that ALCAPA associated with other cardiac defects is often unrecognized before surgery. Pulmonary hypertension due to left-to-right shunt or coarctation can maintain an anterograde flow in the anomalous coronary artery until cardiac repair. Myocardial ischemia will only become apparent once the defect has been repaired when pulmonary pressure lower. In this series postoperative survival was compromised mostly due to complications occurring after complex or repeated cardiac surgery.

http://dx.doi.org/10.1016/j.acvd.2013.06.025

20 The medical past of adults with complex congenital heart disease impacts their social development and professional activity


Advances in surgery and therapeutic catheterization in recent decades increased steady life expectancy and prevalence of adult patients with congenital heart disease (ACHD). We assessed medical and psychosocial parameters of ACHD according to the complexity of the disease.

Methods.--- We included from a single-center prospective observational cohort study which started in January 2013, 68 ACHD patients (40.±13.9 years old, max 77) followed in cardiology unit from January to April 2013 who answer a questionnaire assessing daily activity and psychosocial being. Complexity of the disease was classified according to the 32nd Bethesda conference.

Results.--- Cardiac malformations were simple (S) in 30 patients (44.1%), moderate (M) in 26 (38.2%) and complex (C) in 12 (17.7%). Pediatric cardiac surgery had been performed in 84.2% of M and C
patients. Patients were referred for interventional catheterization (n = 12, 17.6%), for surgery (n = 5, 7.4%), for acute heart failure (n = 3, 4.4%, 100% type C) or for medical follow-up. Complications [stroke (n = 2/12), heart failure (n = 10/12), arrhythmia (n = 7/12), pulmonary hypertension (n = 5/12), death (n = 1/12)] were mainly observed in C patients (P = 0.018). C and M patients had lower education level (respectively 14.3%, 12.5% vs. 55.7% had Baccalaureat, P = 0.019). C patients had less access to driving license (P = 0.02). Only 1/3 of C patients had an active employment. Physical activity was lower in C patients (P = 0.02, none activity in 87.5% but 57% had been previously contra-indicated to sport). There was no significant difference between the three groups regarding antidepressant medication (8.2%, P = 0.578), psychological follow-up (19.0%).

Conclusion.— ACHD patients with complex heart disease have reached adulthood as survivors. Their medical past has impacted their social development and professional activity. Specific attention should be paid to the schooling of children with complex congenital heart disease.

http://dx.doi.org/10.1016/j.acvd.2013.06.026

21 Quality of life among children with congenital heart diseases: Comparative multi-center cross-sectional study

P. Amedro, R. Dorka, S. Moniotte, S. Guillaumont, M.C. Picot, P. Auquier

a Pediatric and Congenital Cardiology, Montpellier University Hospital, France
b EA 3279, Public Health Department, Marseille University of Medicine, Marseille, France
c Pediatric Cardiology, UCL St-Luc, Brussels, Belgium
d Pediatric Cardiology, Institut-St-Pierre, Palavas-Les-Flots, France
e Epidemiology and Biostatistics Department, Montpellier University Hospital, Montpellier, France

Introduction.— Very few comparative studies have yet reported health related quality of life (HRQoL) among children with congenital heart diseases (CHD).

Method.— We prospectively recruited children aged 8 to 18 with CHD (group 1, n = 282) and same age randomized control population selected among schools (group 2, n = 164). CHD were recruited in two tertiary care centers in France and Belgium. Thirty families refused to participate in group 1 and 150 in group 2. Primary outcome was the scoring for each dimension with Kidscreen-52 for children and Kidscreen-27 for parents (French version of the generic validated pediatric HRQoL questionnaire). Secondary outcomes were: severity class of CHD from 1 to 4 (from Uzark), cardiopulmonary exercise test (VO2max, anaerobic threshold, VE/VCO2 slope, oxygen uptake efficiency slope), and PedsQL scoring (non-validated HRQoL generic questionnaire, French and Belgium versions, self and parents reports).

Results.— Two hundred and eighty-two children with CHD (sex ratio 1.9 — mean age 12.3 ± 3) and 164 children in schools (sex ratio 1.1 — mean age 12.8 ± 2.4) were recruited. Both centers were comparable for most demographic and clinical data. In most dimensions self-reported QoL scores among children with CHD were not different from control group, except for physical well-being (mean 46.5 ± 10.2 vs. 49.9 ± 8.6, P < 0.05). QoL in lower severity classes was not significantly different from controls. Children with severe CHD (class 4) had lower QoL in physical well-being (43.34 ± 9.64, P < 0.05). In group 1, parents’ reported QoL was lower than their children’s evaluation in several dimensions (physical, social, school). In control group, scores between self-reported and parents reported QoL were identical. Teenagers had higher QoL scores than younger children. Cardiopulmonary exercise test is well correlated to HRQoL, especially for well-being dimension.

Conclusion.— QoL among children with CHD is close to that of same age healthy children except for physical well-being. HRQoL for low severity CHD patients is similar to controls. QoL for severe CHD is the most impacted. Further studies should evaluate QoL evolution in time per patient.

http://dx.doi.org/10.1016/j.acvd.2013.06.027