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Atrioventricular discordance with ventriculo-arterial concordance: Diagnostic challenge, surgical management and long-term outcome
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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 (D,L,D) segmental arrangement, two had anatomically correct 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. 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.

Anomalous left coronary artery from the pulmonary artery associated with other cardiac defects: A difficult joint diagnosis
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Background. — Anomalous left coronary artery connected to 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 lowers. This series postoperative survival was compromised mostly due to complications occurring after complex or repeated cardiac surgery.

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

Background. — 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 (40.8±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 defects.