Atrioventricular discordance with ventriculo-arterial concordance: Diagnostic challenge, surgical management and long-term outcome
Daniela Lauxa, Lucille Houyelb, Fanny Bajollea, Younes Boudjemeline, Damien Bonnetb
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 (D, S, D) 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 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. In this series postoperative survival was compromised mostly due to complications occurring after complex or repeated cardiac surgery.

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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 (40.2±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