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: Thirteen patients with ALCAPA and other cardiac defects were identified. Five patients had had a prenatal diagnosis of congenital heart disease concerning the associated cardiac defect. The heart defect most frequently encountered in association to ALCAPA was aortic coarctation (n=4) followed by tetralogy of Fallot with or without pulmonary atresia (n=3). There was one case of hypoplastic left heart syndrome, one right aortic arch, one congenital mitral malformation and one infant with divided left atrium and anomalous venous return. Only three patients had a complete diagnosis of the cardiac defect and the left coronary abnormality before surgery. In four 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. Half 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 misdiagnosed before surgery. Pulmonary hypertension due to 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 especially if the coronary anomaly had not been diagnosed preoperatively.