**Surgical repair of Shone’s complex with anomalous origin of the left coronary artery arising from the right pulmonary artery**

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**Abstract**

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly. ALCAPA from the right pulmonary artery (RPA) is a small subset of this anomaly, with only a few reported cases to date. We describe an extremely uncommon association of Shone’s complex with ALCAPA from the RPA in a case where simultaneous surgery for both anomalies was successfully carried out in a neonate.

**Keywords:** Shone complex • Anomalous origin of the left coronary artery from the pulmonary artery • Aortic coarctation • Congenital heart disease • Congenital heart defects • Neonate

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**INTRODUCTION**

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is usually seen as an isolated anomaly in clinical practice, but, in rare cases, it can be associated with other complex congenital heart defects (CHDs). There are some reports of its association with tetralogy of Fallot, transposition of the great arteries and hypoplastic left heart syndrome [1, 2]. The preoperative diagnosis of ALCAPA and other cardiac lesions is very difficult because symptoms of heart failure secondary to myocardial ischaemia may be attributable to an associated CHD, which can change the natural history of ALCAPA. Surgical correction of these associated defects in cases without a detailed coronary artery assessment can also pose a challenge for cardiac surgeons.

**CASE REPORT**

A 1-week old female infant was admitted to the peripheral children’s hospital with tachydyspnoea and peripheral cyanosis after cardiopulmonary resuscitation due to asphyxia. An initial echocardiography showed signs of congestive heart failure and patent ductus arteriosus with possible aortic coarctation. Anti-congestive and prostaglandin therapy were initiated. Following stabilization, the baby was referred to our institution. The electrocardiogram (ECG) revealed significant ST elevation in the anterior and lateral leads. Echocardiography (Fig. 1A and B) identified an anatomy consistent with Shone’s complex. The parachute mitral valve was hypoplastic with mild stenosis (diameter 7.5 mm, mean gradient of 4.2 mmHg, z-score of -2.9), dysplastic aortic valve with severe stenosis (diameter of 4.5 mm, mean gradient of 14.2 mmHg, z-score of -3.8), mildly hypoplastic aortic arch with periductal aortic coarctation. The left ventricle was small compared with the right ventricle, with reduced left ventricular function (EF 34%). The coronary arteries were not examined in detail, but were thought to be normal. Due to the possible existence of an aberrant subclavian artery, cardiac catheterization was performed to confirm the exact aortic arch anatomy. But during the intervention the orifice of the left coronary artery (LCA) at the aortic sinus was not identified. Subsequent pulmonary angiography confirmed the diagnosis of ALCAPA (probably from the main pulmonary artery) (Fig. 1C and D).

Immediate surgery was performed through a median sternotomy. Aortic and bicaval cannulation was performed and a cardio-pulmonary bypass was established. The right and left pulmonary arteries were mobilized and the LCA was identified as originating not from the pulmonary trunk, but from the RPA. The right coronary arterial orifice was in the normal location. Both branches of the pulmonary artery were distally snared to avoid coronary steal and myocardial ischaemia, and the baby was cooled to 17°C. On ECG, a reduction in ST elevation was observed. During this time, the aortic arch was mobilized and the supra-aortic vessels were snared. Prostaglandin infusion was ceased. In deep hypothermic circulatory arrest, the coarctation was resected and the complete aortic arch was enlarged with an autologous pericardial patch (Fig. 2A). After applying an aortic cross-clamp, blood cardioplegia was delivered. The main pulmonary artery was transected for the inspection of the RPA from inside and outside. Inspection of the pulmonary artery revealed that the LCA originated from the distal part of the RPA and descended vertically along the aorta (Fig. 2B). A coronary button of the LCA was excised from the RPA. An additional dose of cardioplegia was given directly in the mobilized LCA. The anomalous coronary artery was reimplanted to the posterior wall of the ascending aorta, after transverse aortotomy...
The defect in the pulmonary artery was reconstructed with an autologous pericardial patch. In addition, the aortic valve was inspected, and a functional bicuspid, dysplastic valve with stenosis of the annulus was found (Fig. 2D). An aortic valvulotomy was then carefully performed. Finally, the aorta and main pulmonary artery were re-anastomosed. Weaning from the bypass was performed without any problems, and no changes in ECG were encountered. The patient was extubated on postoperative day 2. The postoperative echocardiographic images (2 weeks after surgery) demonstrated a good postoperative result. In the parasternal short-axis view, an antegrade flow in the left coronary artery could be seen (E). Suprasternal view showing no re-coarctation stenosis and a normal aortic arch with laminar flow (F). RV: right ventricle; LV: left ventricle; MV: mitral valve; Ao: aorta; AoV: aortic valve; LCA: left coronary artery; RCA: right coronary artery; PA: pulmonary artery.

**COMMENT**

ALCAPA is a rare congenital coronary anomaly responsible for cardiac failure due to myocardial ischaemia with a high mortality rate in infants. Its incidence and pathophysiological mechanisms, when associated with other CHDs, are unknown.

The relationships between left ventricular hypoplasia and several coronary anomalies have been documented in patients with hypoplastic left heart syndrome (HLHS), with the most common being coronary fistulas or ventricular–coronary connections [3]. In our case, the patient did not have any coronary fistulas or connections from the LCA to the cardiac cavities or to the RCA. The LCA was separately developed from other structures of the
heart and had no ostial stenosis, which could be present in such cases.

An exact preoperative diagnosis in cases of ALCAPA associated with other CHDs is very difficult, mostly due to specific haemodynamics masking myocardial ischaemia. The presence of neonatal isthmic stenosis can also explain the development of cardiac failure with left ventricular dysfunction and is often misinterpreted. Preoperative recognition of ALCAPA is critical for successful management of cardiopulmonary bypass and the surgical strategy.

In most cases with a combination of ALCAPA and other cardiac lesions, the presence of ALCAPA was not diagnosed preoperatively, leading to poor results. Nathan et al. described 6 cases of ALCAPA in HLHS [2]. Diagnosis was made preoperatively by echocardiography and catheterization in only 1 patient. In 5 patients, the diagnosis was made intraoperatively, in 4 of these patients late in the procedure. All of these 4 patients had inadequate coronary perfusion and died of myocardial failure in the postoperative period. Saroli et al. reported 3 patients with hypoplastic left heart syndrome and a case similar to ours with coronary anomalies [4]. A diagnosis was made intraoperatively in 2 of these patients, resulting in coronary injury with a lethal outcome. In their review of cases of ALCAPA with other cardiac defects in 12 patients, Laux et al. [1] reported that only 4 patients had a complete diagnosis before surgery. In 6 patients, a diagnosis was made after cardiac repair. The postoperative mortality rate in these 12 patients was 58% (7 patients). Ilic et al. described an infant with ALCAPA associated with Scimitar syndrome and aortic coarctation. In this patient, ALCAPA was diagnosed postoperatively after coarctation repair [5].

Adequate cardiopulmonary bypass management of these patients is very important, and careful examination of the coronary anatomy before initiation of cardiopulmonary bypass is essential. Double cannulation of the aorta and the pulmonary artery (for LCA perfusion) can also represent an option. Ductal patency with clamping of pulmonary artery branches can prevent coronary steal and myocardial ischaemia. Prostaglandin infusion must be continued until completion of the ductus arteriosus ligation. Cardioplegia can be injected simultaneously in the aortic root and isolated pulmonary artery, or in the mobilized coronary artery.

In conclusion, we present a neonate with a very rare combination of Shone’s complex and ALCAPA from the RPA. Surgical correction of both congenital anomalies was performed simultaneously with an excellent outcome. For successful management of these patients, preoperative identification of the coronary artery anatomy is critical, which might necessitate a combination of imaging strategies.

Conflict of interest: none declared.

REFERENCES


