Surgical correction of a congenital coronary arterial fistula and a massive sinus of Valsalva aneurysm

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Abstract
The case of a 22-year old male patient who presented with a congenital coronary arterial fistula from the right coronary artery to the right ventricle is described. The arterial fistula had led to an aneurysm of the right sinus of Valsalva and the proximal right coronary artery. The aneurysm was incised from the aorta and the fistula closed with a pericardial patch. The diameter of the aneurysm was reduced by plication. Successful occlusion of the fistula and the competence of the aortic valve were confirmed by transoesophageal echocardiography.

Keywords: Acyanotic congenital

INTRODUCTION
Congenital coronary arterial fistulas are the most common haemodynamically significant congenital coronary abnormalities. Their incidence is reported to be 0.002% in the general population [1] and to account for ~0.2–0.4% of all congenital cardiac anomalies [2]. These malformations may lead to cardiac failure, infectious endocarditis, arrhythmia and myocardial hypoperfusion [3]. In the present case, the specific danger of developing aortic regurgitation and dissection/rupture had to be considered. Percutaneous closure techniques have been introduced into clinical practice, thus avoiding the risk and complications of sternotomy and cardiopulmonary bypass [4]. However, many lesions cannot be treated by a transcatheter approach. In our case, the fistula could potentially have been closed interventionally, but the massive dilatation of the right sinus of Valsalva and the proximal part of the right coronary artery (RCA) and the extension of the RCA beyond the aneurysm mandated surgical restoration.

CASE REPORT
A 22-year old male patient (186 cm, 78.7 kg, BMI 21.7) experienced palpitations at rest. Echocardiography revealed a congenital coronary arterial fistula. On admission, auscultation revealed a continuous 3/6 systolic-diastolic murmur in the third and fourth intercostal space, parasternal to the left. The electrocardiogram was normal. On echocardiography, severe left ventricular (LV) dilatation (left ventricular internal dimension at diastole 72 mm) was seen, LV ejection fraction was slightly reduced and RV function was normal. The proximal RCA (diameter 35 mm) (Fig. 1a) and the right sinus of Valsalva were massively enlarged. A systolic-diastolic colour-flow signal was seen leading into the proximal right ventricular outflow tract (RVOT) (flow 2.5 m/s, Pmean 12 mmHg) (Fig. 1b). The aortic valve was competent. The coronary computed tomography revealed the enlarged bulbus aortae (52 mm) due to the aneurysm of the right sinus Valsalva. RVOT and truncus pulmonalis (40 mm) were dilated.

Upon operation, a median sternotomy was performed and CPB started after bicaval venous cannulation with moderate systemic hypothermia (32°C). For myocardial protection, we used 1500 ml of Bretschneider solution infused into the aortic root. After incision of the ascending aorta, 1000 ml of Bretschneider solution was applied through direct coronary ostial infusion into the left coronary artery. The right atrium was incised and the opening of the fistula on the ventral aspect of the RV inspected via the tricuspid valve. The ostium of the fistula measured ~8–10 mm, but sufficient exposure of the transatrial closure could not be achieved due to the cranial position, hypertrophic myocardium and the proximity of the tricuspid valve. Therefore, a longitudinal aortotomy was performed and the incision extended into the large aneurysm and the proximal RCA until the ostium of the fistula into the RV was well exposed (Fig. 2a). The aneurysm continued into a miniture RCA of ~1 mm in diameter (Fig. 2b). The ostium of the fistula was closed by a circular pericardial patch using a running 5-0 Prolene suture (Fig. 2c). The wall of the aneurysm was partly resected (Fig. 2d) and plicated to further reduce its size. The weaning of the CPB was uneventful. A postoperative transthoracic echocardiography and angiography revealed the complete closure of the arterio-venous fistula, and a competent aortic valve. The diameter of the right sinus of...
Valsalva was reduced from 63.96 to 28.43 mm. The aortic root measured 49.37 mm postoperatively.

**DISCUSSION**

Congenital coronary arterial fistulas may cause late complications such as myocardial infarction, endocarditis and death. Liberthson et al. reviewed 174 cases and found a related complication rate of 35% in adults (≥20 years). Death occurred in 14%. Spontaneous closure of coronary fistulas is extremely rare. Surgical closure is effective, safe and provides excellent long-term results [5]. Experience with percutaneous interventional closure devices is limited, but may be considered as an option in carefully selected patients [2, 4]. In the present case, reduction of the massive sinus and coronary artery aneurysm and preservation of the extention of the small RCA beyond the aneurysm mandated surgical therapy. An alternative procedure would have been aneurysm resection and patch reconstruction of the sinus of Valsalva followed by patch closure of the fistula. However, this would have required a right mammary artery bypass to the distal RCA, which was much too small for this approach.

**Conflict of interest:** none declared.

**REFERENCES**