Unusual aberrancy of right coronary artery (RCA) in patient with double-outlet right ventricle (DORV) with pulmonary stenosis (PS)

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Unusual aberrancy of RCA

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Abstract
The incidence of coronary artery anomalies in double-outlet right ventricle (DORV) or tetralogy of Fallot (TOF) has been reported to be 2% to 10%. Surgical correction in such a scenario, across a narrowed and under-developed right ventricular outflow tract (RVOT), poses a significant challenge and has been associated with increased morbidity and mortality. These combinations at times may change the approach of primary repair to one of initial palliation. Single coronary artery, meaning anomalous origin of the right coronary artery from the left anterior descending artery, is a benign and very rare coronary artery anomaly. Here, we report the case of an 11 year-old boy with double-outlet right ventricle (DORV) with pulmonary stenosis (PS) where pre-operative catheterisation and multi detector CT angiography (MDCT) revealed anomalous right coronary artery arising from left anterior descending artery.

Key words: coronary artery anomalies; double-outlet right ventricle; tetralogy of Fallot; anomalous right coronary artery; multi-detector CT angiography

Introduction
Anomalous right coronary artery (RCA) usually arises from the left sinus itself, or directly from the left main coronary artery or its vicinity. Coronary circulation from a single coronary artery usually has little clinical significance; however, variability in anatomical origin and course can give rise to a malignant course resulting in sudden cardiac death and an increased risk of ischaemia. Anomalous origin of RCA from left anterior descending (LAD) has rarely been reported in the literature [1]. Most of these cases have been reported in a structurally normal heart, and rarely in patients with double-outlet right ventricle (DORV) or tetralogy of Fallot [2, 3]. Therefore, they have been identified mostly as an incidental finding, and rarely present in patients with congenital cyanotic heart disease.

Case report

An 11 year-old boy presented with central cyanosis with pandigital clubbing for evaluation. His blood pressure was 100/70 mm Hg in right upper limb in supine position and pulse rate was 78/min, regular, normal volume with no radio-radial or radio-femoral delay, and with all peripheral pulses palpable. There was a history of squatting in the childhood. On examination, precordium was silent. On auscultation, S1 was soft, S2 was loud and single, grade 2/6 basal ejection murmur, and continuous murmur in right axilla and subscapular region. Electrocardiogram showed right axis deviation, right atrial enlargement, right ventricular (RV) hypertrophy, and first degree heart block. Chest X-ray showed normal cardiac size, right ventricular type of apex, and oligemic lung field. Therefore, cyanosis with decreased pulmonary blood flow was suspected. Echocardiography demonstrated origin of both great arteries from the anterior RV, the absence of left ventricular outflow other than ventricular septal defect (VSD), and the discontinuity of mitral and semilunar valve. Aorta was overriding the ventricular septum by more than 50%. There was pulmonary stenosis with $V_{\text{max}}/\Delta P_{\text{max}}$ of 4.96/98 (Figure 1). Cardiac catheterisation was planned after proper consent to rule out any additional muscular VSD, major aorto pulmonary collaterals (MAPCA), and coronary artery anomalies. Cannulation of left main coronary artery showed its normal course dividing into left anterior descending artery (LAD) and left circumflex artery (LCx). However, right coronary artery (RCA) was originating from proximal LAD before the origin of first major septal branch (Figure 2). On aortic root angiogram using a pigtail catheter, RCA could not be visualised from the right aortic sinus. All three arteries appeared free of disease. To confirm the anomalous origin, and to better delineate the course of the artery and
find major aorto-pulmonary collaterals, 128-slice multi-detector computed tomography (MDCT) coronary angiography was performed. This revealed single left coronary artery and RCA originating from the proximal LAD, coursing in front of the right ventricular outflow tract, and travelling down the right atrioventricular groove. Therefore, left main coronary artery was functionally trifurcating into LAD, LCx and RCA (Figures 3, 4). The patient was referred for surgical correction.

Discussion

In most previous reports of anomalous RCA arising from LAD, it has been described as a single anomalous vessel originating after the first septal perforator of the LAD, which courses anteriorly to the right ventricular outflow tract to reach territory normally served by the RCA [4]. An anomalous RCA carries a different meaning and perspective for surgeons because its location interferes with the usual surgical repair of an obstructed RVOT, and its sacrifice can seriously compromise the viability of a significant portion of the myocardium [5].

In such situations, the ventriculotomy needs to be displaced, to either the left or the right, to avoid damage to the anomalous coronary artery. Furthermore, the ventriculotomy will be oblique rather than vertical. Surgical correction of congenital heart disease in the past three decades has witnessed enormous strides as early primary repair for the vast majority of cardiac malformations is now carried out rather than initial palliation. This started with tetralogy of Fallot (TOF), and is now also being performed for other complex lesions such as DORV, with excellent results [6]. It establishes normal anatomic and physiologic conditions as early in life as possible. This avoids the undesirable long-term effects of unrepaired congenital heart disease, as well as the complications of palliative operations and the risk of multiple surgeries [7]. Despite this, the presence of an anomalous coronary artery crossing an obstructed outflow tract is traditionally considered to be a contraindication to primary repair in infancy. This is because of the morbidity and mortality associated with this condition during complete cardiac repair when one needs to enlarge the right ventricular outflow tract (RVOT) which may injure the anomalous vessel [8–10].

Also, anomalous RCA, when it travels across the lower portion of the RVOT, may hinder the full mobilisation of the main pulmonary artery (MPA) and its confluent branches [3]. In our case, anomalous RCA was arising from proximal LAD, rather than an extension of septal perforator. In such cases, a reverse pulmonary artery flap repair over the anomalous coronary, a transannular patch repair under a mobilised LAD, or a displaced ventriculotomy
and sub coronary suture lines are alternatives that should be considered [3]. Finally, a conduit may be required as a last resort to allow primary repair.

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References


**Figure 1.** 2D transthoracic echocardiography showing classical features of double-outlet right ventricle with pulmonary stenosis: A. Ventricular septal defect with aortic override; B. Aorto-mitral discontinuity; C. Pulmonary stenosis

**Figure 2.** Left main trunk dividing into LAD, and LCx with RCA (white arrow) arising from proximal LAD: A. Left anterior oblique cranial view; B. Straight lateral view

**Figure 3.** Multi-detector computed tomography coronary angiography showing RCA (white arrow) arising from proximal LAD

**Figure 4.** Volume rendered reconstruction of coronary computed tomography angiography showing RCA (white arrow) arising from proximal LAD coursing in front of right ventricular; blue arrow — major aorto-pulmonary collaterals