Anomalous Course of the Left Main Coronary Artery in Tetralogy of Fallot

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An extremely rare coronary artery anomaly where the left main coronary artery arose anteriorly from the right coronary sinus and coursed in front of the right ventricular outflow tract was present in a patient with tetralogy of Fallot. Preoperative angiography was interpreted as normal. Operative recognition was prevented by dense adhesions and a partial intramural course. Division of the vessel at repair resulted in death of the patient. The angiographic pattern associated with this anomaly is very unusual, and in many views looks deceptively normal. Details are presented.


Surgically important coronary artery anomalies may coexist with tetralogy of Fallot. Fellows and associates [1], in 1975, described five known abnormal coronary patterns. In three of these, an important coronary artery traversed the right ventricular outflow tract (RVOT) that would be at risk for division during a repair employing a right ventriculotomy. Other groups have confirmed the occurrence of these anomalies in 2% to 12% of patients [2–5]. Although the most common of these arrangements is the left anterior descending coronary artery (LAD) arising from the right coronary artery (RCA), variations of single left or right coronary artery, as well as paired LAD, may cross the RVOT. The existence of any of these surgically important anomalies described to date should, therefore, be ruled out by observing an RCA and a single LAD arising from the left main coronary (LMCA). Recommendations have been made regarding the cineangiographic views most likely to define these anomalies [6, 7].

We recently encountered a situation where preoperative cineangiography clearly documented a separate RCA and a single LAD arising from the LMCA. Mortality nonetheless resulted from coronary artery division at the time of operation. In this patient, a long LMCA arose as a separate vessel from the right coronary cusp, coursed anteriorly and high over the RVOT, and then divided into a circumflex artery (Circ) and an LAD. A combination of dense pericardial adhesions from a previous operation and a partial intramural coronary artery course prevented recognition of this anomaly, and the LMCA was divided in the course of the RVOT incision. We present this case to emphasize the existence of this extremely rare coronary artery anomaly in the presence of tetralogy of Fallot and to suggest the cineangiographic findings that help to identify it.

A 3.5-kg female patient, born at 40 weeks’ gestation to a gravida 1, para 0-1 mother who had received prenatal care, was recognized early to be cyanotic. A diaphragmatic hernia and tetralogy of Fallot with marked pulmonary artery hypoplasia were documented. Repair of the diaphragmatic hernia was followed by a palliative central shunt. At the initial palliation, a prominent vessel was noted arising from the right coronary sinus, which appeared to course toward the RVOT. Thus, suspicion was raised regarding the presence of an aberrant LAD arising from the RCA. Subsequent palliation consisted of a balloon pulmonary valvulotomy.

Evaluation for definitive repair at 10 months of age revealed systemic pressures throughout the entire right heart and pulmonary vessels except in the distal right pulmonary artery, where the pressure was 75% systemic. There was concern regarding multiple distal stenoses. Transplant options were discussed. Despite the high risk, a corrective operation was elected.

Multiple angiograms were available for coronary artery evaluation. It was obvious that she did not have an aberrant LAD from the RCA as suspected initially because the LAD arising from the LMCA was clearly seen (Fig 1). Thus, the suspicion noted from the original operation was lessened. It was also clear that there were two separate coronary arteries. Therefore, an operation was planned whereby the RVOT would be opened and an orthotopic homograft placed to more effectively deal with the elevated pulmonary resistance.

At operation, the adhesions from the previous operation were dense and completely obscured the anterior...
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Comment

Although previously described in association with normal hearts, this particular coronary anatomy has not been generally recognized in association with tetralogy of Fallot [8–10]. However, this anomaly must be recognized because of its surgical significance. The angiographic identification of the courses of the LMCA, LAD, and RCA is sometimes difficult because of varying degrees of right heart hypoplasia and the obliquity of the standard angiographic projections. In the typical coronary artery anomaly in tetralogy of Fallot, the LAD arises from the RCA and courses in front of the RVOT. The Circ originates separately from the left coronary sinus and courses directly posteriorly and toward the apex. In this case, the LMCA originated from the right coronary sinus anteriorly and coursed in front of the RVOT, and the Circ wrapped around the left lateral aspect of the pulmonary root to the posterior aspect of the heart (see Fig 1). This is a distinctly abnormal finding and has only been described with an anomalous LMCA course.

Therefore, we recommend that, in addition to identifying two separate orifices to the LMCA and RCA, coronary angiography must delineate the course of the Circ. If standard projections are not adequate, multiple projections including four-chamber or steep caudal with left oblique angulation may be necessary. In addition to identifying the far more common coronary artery anomalies, recognition of an anomalous LMCA by identifying

surface anatomy of the heart. As the ventriculotomy was made, a coronary artery was transected deep beneath the cardiac surface. Because we believed that the LAD did not aberrantly arise from the RCA, we proceeded with the repair, assuming that a conal branch had been transected. After release of the cross-clamp, myocardial activity initially returned but did not support separation from bypass. Internal mammary artery grafting did not appear feasible because of the small size of the patient and the distance that would have needed bridging secondary to the presence of the orthotopic graft. Autopsy revealed that the LMCA arose from the right coronary sinus as a completely separate vessel (Fig 2) that coursed anteriorly across the RVOT, where it had been divided, continued to the septum, and then divided into an LAD and a Circ. The vessel coursed 5 mm below the cardiac surface.

Fig 1. (A) Anterior/posterior aortogram with the circumflex (Circ) and left anterior descending (LAD) arteries bifurcating from the left main coronary artery (LMCA) lateral to the pulmonary root. (B) The curved arrows in the lateral projection show the posterior course of the Circ after bifurcating with the LAD from the anteriorly positioned LMCA. (RCA = right coronary artery.)

Fig 2. View of the right coronary sinus from behind looking toward the anterior aspect of the root. The straight arrow points toward the orifice of the right coronary artery and the curved arrow toward the orifice of the left main coronary artery. (LCC = left coronary cusp; NCC = noncoronary cusp; RCC = right coronary cusp.)
a Circ wrapping around the pulmonary root will allow appropriate surgical planning in this exceedingly rare anomaly in tetralogy of Fallot. Whether echocardiography can exclude this variant in previously operated patients, when surface anatomy is potentially obscured, remains to be seen.

References


Late Contralateral Lobectomy After Single-Lung Transplantation for Emphysema

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Cases of hyperinflation of native emphysematous lung have been reported in the early period after single-lung transplantation. We report a case of a similar complication that occurred 2 years after transplantation and was successfully treated by lobectomy.


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Single-lung transplantation (SLT) has been regarded as contraindicated in chronic obstructive pulmonary disease. Ventilation-perfusion imbalance and graft compression by the distended emphysematous native lung (NL) were considered obstacles to SLT. Cases of graft compression by the NL in the early phase after intervention have been reported in the literature. We report a case of late graft compression that occurred 2 years after transplantation and was successfully treated by lobectomy.

The patient was 59 years old and had had a panlobular emphysema since he was 18 years old. Emphysema increased with smoking (40 pack-years) until severe respiratory insufficiency developed. The forced expiratory volume in 1 second was 0.35 L (11% of the predictive value). The patient was confined to bed with continuous oxygen therapy (1.5 L/min). Blood gases without oxygen were as follows: arterial oxygen tension, 38 mm Hg; arterial carbon dioxide tension, 48 mm Hg. Single-lung transplantation was performed on the left lung. The postoperative period was uneventful. Two months after intervention, the clinical state was good without oxygen therapy, oxygen and carbon dioxide tension improved to 91 and 36 mm Hg before effort and 93 and 31 mm Hg after effort (25 W during 5 minutes). The forced expiratory volume in 1 second was 2 L, 62% of the predicted value.

Eighteen months after transplantation, the patient was dyspneic again. Transbronchial biopsy found a bronchiolar fibrosis of the left lower lobe. Oxygen and carbon dioxide tension were 86 and 38 mm Hg, decreasing to 77 and 40 mm Hg after effort (Fig 1). The forced expiratory volume in 1 second was 1.2 L, 36% of the predicted value. Hyperexpansion of the remaining recipient's lung was found on chest roentgenogram (Fig 2). Twenty-five months later, the patient was not able to walk more than 500 m at a slow pace, and he needed two pauses to climb up one floor. Oxygen and carbon dioxide tensions were 84 and 40 mm Hg, changing to 73 and 40 mm Hg after effort, and the forced expiratory volume in 1 second was...