Tetralogy of Fallot With Dextrocardia and Anomalous Coronary Artery: A Case Report

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DOI: 10.1177/2150135120911342

Abstract

We report a rare case of tetralogy of Fallot with dextrocardia and anomalous coronary artery. Although this is an unusual complex disease, we have successfully repaired it with a combination of transatrial/transpulmonary and trans–right ventricle techniques.

Introduction

Tetralogy of Fallot (TOF) is a frequently diagnosed congenital heart disease. Common concomitant defects include patent foramen ovale and patent ductus arteriosus. Anomalous origin and course of a major coronary artery such that an important branch traverses the right ventricular outflow tract, on the other hand, is an infrequent anomaly associated with TOF, which occurs in 2% to 9% of TOF cases. Tetralogy of Fallot with dextrocardia is a rarely seen pathology with a rate of 1.4%. To the best of our knowledge, so far there has been one case reported since 1965 of TOF associated with both anomalous major coronary branch and dextrocardia, that case having been encountered in Japan. Accordingly, we report here a case with TOF with dextrocardia and anomalous coronary artery which was successfully repaired.

Clinical Case

A seven-month-old girl (weight: 7 kg, height: 80 cm, body surface area: 0.39 m²) presented to us for evaluation and treatment of TOF, which was first diagnosed at age five months. Her main symptom at admission was mild cyanosis of the lips, and her vital signs were as follows: SpO₂ of 84% (measured at fingertip), heart rate of 120 beats per minute, and blood pressure of 85/50 mm Hg. Chest auscultation revealed a grade 3/6 systolic murmur, best heard at the third intercostal space, adjacent to the left border of the sternum. Laboratory results showed that hemoglobin was 15.9 g/dL, hematocrit was 48.2%, and serum biochemistry was within normal range. Her chest X-ray showed right-deviated cardiac silhouette (Figure 1).

Abdominal ultrasound revealed situs inversus of the viscera. Doppler echocardiography confirmed the diagnosis of dextrocardia and revealed a perimembranous ventricular septal defect (VSD) with a diameter of 11 mm, an overriding aorta by 50%, and right ventricular outflow tract obstruction with a pressure gradient across the right ventricular outflow tract (RVOT) and pulmonary valve of 60 mm Hg. The diameter of pulmonary valvular annulus was 9 mm. The sizes of the right and left pulmonary arteries were 6 and 5 mm, respectively (Figure 2).

The patient underwent surgical repair. After pericardiotomy, we observed that the right coronary artery (RCA) originated from the left anterior descending artery (LAD) and crossed the infundibulum (Figure 3 and Supplemental Video).

Submitted October 2, 2019; Accepted February 17, 2020.

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We performed surgical repair utilizing a combined approach including both a transatrial/transpulmonary approach to resect RVOT musculature causing outflow obstruction while preserving the pulmonary valvular annulus and a trans–right ventricular approach (limited ventriculotomy placed caudally and parallel to the course of the crossing RCA), which in the unusual setting of dextrocardia and mirror image anatomy enabled us to confidently address the VSD, while minimizing the risk of injury to the conduction system or the tricuspid valve and its tensor apparatus. The VSD was closed with a XenoSure patch (LeMaitre Vascular, Inc, MA, USA). The duration of cardiopulmonary bypass was 132 minutes, and the aortic cross-clamp time was 113 minutes. The pressure gradient across the pulmonary valve after successful weaning from cardiopulmonary bypass was 12 mm Hg. The postoperative period was uneventful and ventilation time was 18 hours. The patient was discharged on postoperative day 10 and the echocardiography prior to discharge showed that the VSD was completely closed with no residual shunt. The pulmonary valve was functioning normally with no regurgitation, and the peak pressure gradient across the RVOT was 24 mm Hg.

Discussions

Tetralogy of Fallot accounts for approximately 3.5% of all congenital heart diseases. The frequency of a major coronary artery anomaly in TOF varies from 2% to 9% among studies. This abnormality can be detected preoperatively through echocardiography and angiography, during surgery, or at autopsy. Different variations include LAD originating from the RCA, single RCA ostium, RCA originating from the LAD, single left coronary artery (LCA) ostium, major branches of RCA crossing the infundibulum, and other anomalies such as RCA-to-right ventricle fistula, left circumflex artery originating from the RCA, and so on. Repair techniques in the setting of a major coronary branch crossing the RVOT include approach through the right atrium and pulmonary artery, use of a conduit to connect pulmonary artery and RVOT, and limited right ventriculotomy (with or without mobilization of the crossing coronary artery). The technique of choice depends on the size of pulmonary valvular annulus and the location of the anomalous coronary artery. But transatrial/transpulmonary approach is a generally preferred technique that is used whenever possible.

Tetralogy of Fallot with concomitant dextrocardia in the setting of atriovisceral situs inversus is extremely rare. This combination was first described in 1952 by Scragg and Denny, but there are only few reports on this rare defect. Among 147 TOF adult patients, there were 2 (1.4%) patients with dextrocardia. The altered anatomical location and orientation of the heart typically makes it necessary for the surgeon to carry out the repair from the patient’s left side, including visualization and closure of the VSD as well as relief of RVOT obstruction. This atypical arrangement may make it more difficult to repair the defects (VSD closure, RVOT, and pulmonary artery extension). Consequently, the risk of residual VSD as well as RVOT obstruction and damage to the conduction system may also be higher.

Our patient had a combination of TOF, dextrocardia, and RCA abnormally originating from the LAD. The surgeon must approach the defect from the left of the patient. The total surgical repair was performed using a combination of approaches: through the right atrium, through the pulmonary artery, and through a limited right ventricular opening placed inferiorly and parallel to the RCA where it crossed the infundibulum. We combined these approaches to allow the surgeon to better expose and control the margins of VSD, reducing the likelihood of residual VSD while avoiding injury to the atrioventricular node and penetrating bundle. This approach also makes the RVOT extension easier to better achieve adequate relief of subvalvar RVOT obstruction.

Authors’ Note

Parental consent was provided for publication of this case. Do Anh Tien and Tran-Thuy Nguyen equally contributed to the article.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

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Supplemental Material

Supplemental material for this article is available online.

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