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Root Translocation in Congenitally Corrected Transposition of the Great Arteries with Ventricular Septal Defect and Pulmonary Stenosis, and Other Lesions

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The pulmonary root translocation (PRT) procedure has been used to correct ventriculoarterial discordance or malposition of great arteries since 1994. It was part of the surgical repair of 62 consecutive patients presenting with congenitally corrected transposition of the great arteries (CCTGA) with ventricular septal defect (VSD) and pulmonary stenosis (PS), or other complex congenital heart disease with malposition of the great arteries, VSD, and PS. PRT was performed as follows: removal of the pulmonary artery (PA) with the pulmonary valve from its abnormal position, closure of the consequent hole with an autologous pericardial patch, resection of some conal septum, creation of an intraventricular tunnel connecting the left ventricle to the aorta, and construction of a new right ventricular outflow tract using the translocated PA. In patients presenting with important pulmonary valve stenosis, the pulmonary artery was enlarged with a monocusp valve pericardial patch. The Senning procedure was used with some modification to complete the anatomical repair in CCTGA patients. Overall in-hospital and long-term mortality were 4.8% and 3.4%, respectively. PRT appears to be a good surgical alternative for patients presenting with CCTGA with VSD and PS, and other lesions involving malposition of the great arteries, VSD, and PS.
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Introduction

In 1990, Ilbawi and coworkers[1] were the first to publish the anatomical repair of congenitally corrected transposition of the great arteries (CCTGA). They reported on two patients who were repaired with a combination of the atrial switch (Mustard technique) and the Rastelli procedure. In the long term, the majority of the patients submitted to this approach show good left and right ventricular function, and no, or mild tricuspid and mitral insufficiency, but require various reoperations over time.[2]

In 2004, we began to use pulmonary root translocation (PRT) as part of the anatomical repair of CCTGA, ventricular septal defect (VSD), and left ventricle outflow tract obstruction (LVOTO).[3,4] This was an extension of our routine approach to patients presenting with transposition of great arteries, VSD, and LVOTO, or double outlet right ventricle, malposition of great arteries, VSD, and pulmonary stenosis (PS),[5] which was initiated in 1994. This approach, which preserves pulmonary valve tissues, was designed to reduce the need for reinterventions related to the pulmonary insufficiency or stenosis that often occurs during long-term follow-up from the available techniques that discard the native pulmonary valve.[6–9]

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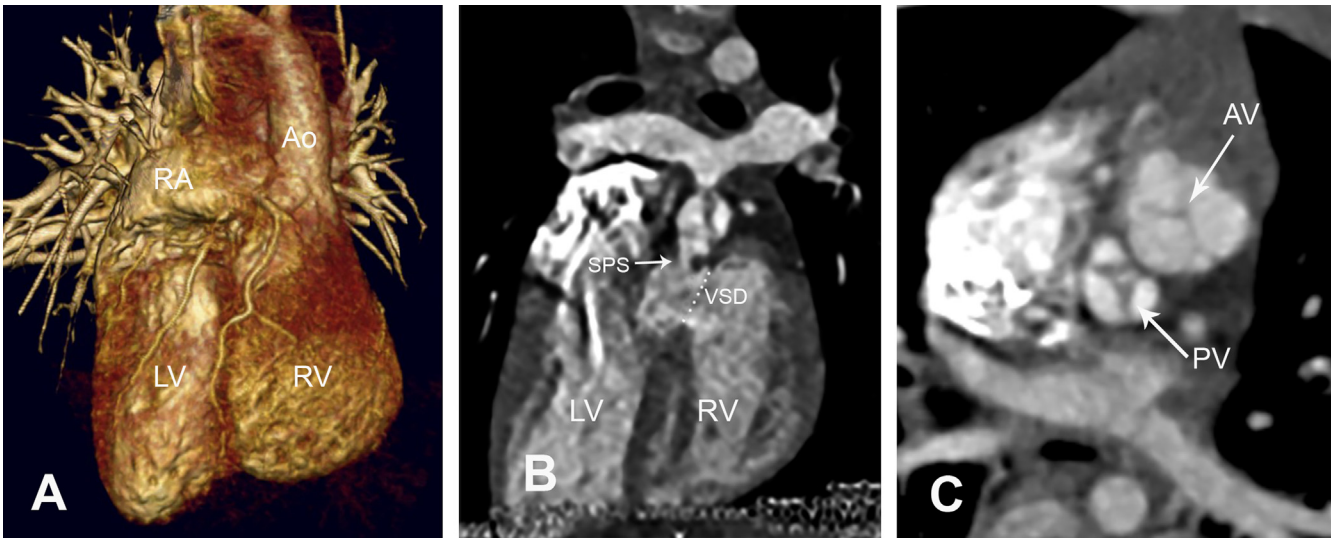

Figure 1 Preoperative computed tomography angiogram of a CCTGA patient showing the right ventricle connected to the aorta (ventriculoarterial discordance), the right atrium connected to the left ventricle (atrioventricular discordance) (A) in association with VSD, sub-pulmonary stenosis (B), and pulmonary valve stenosis caused by thickening of the valve cusps and small pulmonary valve diameter (C). AV, aortic valve; PV, pulmonary valve; SPS, sub-pulmonary stenosis; VSD, ventricular septal defect; LV, left ventricle; RV, right ventricle; RA, right atrium; Ao, aorta.

PRT was used to correct either ventriculoarterial discordance or malposition of the great arteries and consisted of: removal of the pulmonary artery with the pulmonary valve from the left ventricle, closure of the consequent hole in the left ventricle using a glutaraldehyde-treated autologous pericardial patch, resection of some conal septum, closure of the VSD with creation of an intraventricular tunnel to divert blood flow from the left ventricle to the aorta, and anterior pulmonary artery translocation to construct a new right ventricle outflow tract. The atrial switch with the Senning technique with some modification was used to complete the anatomical repair of CCTGA.

Surgical Technique

The operation is performed via median sternotomy. Cardiopulmonary bypass is instituted with aortic and bicaval cannulation. For myocardial protection, we use systemic hypothermia (23 to 25°C) and cold antegrade blood cardioplegia (20 mL/kg), followed by subsequent doses (10 to 15 mL/kg) at 20- to 30-minute intervals during the cross-clamp period. The heart anatomy and the main surgical steps for PRT in CCTGA are depicted in Figures 1 through 4. The postoperative morphological result in the same patient is shown in Figure 5.

The surgical details involved in the use of PRT to repair other lesions, such as dextro-TGA and double outlet right ventricle in association with VSD and PS, have been described elsewhere.[3–5] In patients presenting with important pulmonary valve stenosis (pulmonary valve diameter Z score $\leq$-3 at preoperative echocardiogram or intraoperative right ventricle – pulmonary artery systolic gradient superior to 35 mmHg, after repair), the pulmonary artery was enlarged with a monocusp valve pericardial patch.

From March 2004 to February 2014, we operated on eight patients with CCTGA, VSD, and PS, using PRT as part of the corrective procedure. These patients were grouped with 54 patients that had other cardiac lesions, but also had malposition of the great arteries, VSD, and PS (Table 1), and had undergone PRT since 1994.

Results

Early and long-term mortality are depicted in Table 1. Atrioventricular blockage occurred in three patients: two instances were caused by the surgical procedure, and one patient already had a heart block when referred for surgery.

The two long-term deaths were both sudden deaths. One case was probably related to cardiac arrhythmia in the CCTGA patient; the other may have been related to progressive right ventricular outflow tract (RVOT) obstruction in a dextro-TGA child who was being scheduled for surgical reintervention.

Six patients required surgical reintervention for the following reasons: residual VSD in three patients, LVOTO in one patient, RVOT aneurysm in one patient, and pulmonary valve endocarditis in one patient. Additionally, two patients required pulmonary valve balloon dilation.

Comments

The PRT technique was aimed at maintaining pulmonary valve function as much as possible, including the capacity for growth, as an attempt to avoid the problems inherent in a right ventricle to pulmonary artery conduit. In addition, this technique leaves the aorta untouched in its original anterior