A 34-Year-Old Thai Man Presenting with Pulmonary Stenosis and Heart Failure 24 Years After Surgical Correction with the Rastelli Procedure for Congenital Dextro-Transposition of the Great Artery, Ventricular Septal Defect, and Pulmonary Atresia

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Financial support: None declared
Conflict of interest: None declared

Patient: Male, 34-year-old
Final Diagnosis: Pulmonary stenosis and heart failure 24 years after surgical correction with the Rastelli procedure for congenital D-TGA, VSD, and pulmonary atresia
Symptoms: Dyspnea on exertion
Medication: —
Clinical Procedure: Pulmonary artery plasty • Rastelli procedure • Right-ventricle-to-pulmonary-artery conduit • VSD closure
Specialty: Anatomy • Cardiac Surgery • Cardiology • General and Internal Medicine
Objective: Congenital defects/diseases

Background: Dextro-transposition of the great arteries (D-TGA) with a ventricular septal defect (VSD) and pulmonary atresia is an uncommon congenital conotruncal abnormality. Surgical correction is performed using the Rastelli procedure, which includes a ventricular septal patch to direct blood from the left ventricle to the aorta and a valved conduit to connect the right ventricle to the pulmonary artery. This report is of a 34-year-old Thai man who presented with pulmonary stenosis and heart failure 24 years after surgical correction with the Rastelli procedure for congenital D-TGA, VSD, and pulmonary atresia.

Case Report: A 34-year-old Thai man presented with dyspnea on moderate exertion. His cardiovascular examination revealed a median sternal surgical scar, parasternal heaving, a grade III systolic ejection murmur at the left upper parasternal border, and a single second heart sound. Echocardiography demonstrated degenerative calcification of a severely stenosed pulmonary valve and impaired right ventricular function. A color Doppler M-mode echocardiogram showed VSD patch leakage. A computed tomography scan with 3-dimensional heart reconstruction demonstrated a significantly stenosed branch pulmonary artery. Right and left heart catheterization confirmed the multi-site stenoses were hemodynamically significant. The patient underwent surgery for VSD closure, placement of a right-ventricle-to-pulmonary-artery conduit with a polytetrafluoroethylene graft, and pulmonary artery plasty to correct the stenosis at the branch of the pulmonary artery.

Conclusions: The long-term complications of the Rastelli-type operation seen for D-TGA with a VSD and pulmonary atresia included a right-ventricle-to-pulmonary-artery conduit obstruction, VSD patch leakage, and re-stenosis of the peripheral pulmonary stenosis. Multimodal imaging was informative in planning for reoperation.

Keywords: Arterial Switch Operation • Pulmonary Atresia With Ventricular Septal Defect • Stenosis, Pulmonary Artery • Transposition of Great Vessels • Cardiac Surgical Procedures • Multimodal Imaging • Echocardiography, Doppler • Imaging, Three-Dimensional • Cardiac Catheterization • Pulmonary Valve Stenosis • Case Reports • Heart Failure

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/933078

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Background

Dextro-transposition of the great arteries (D-TGA) is characterized by atrioventricular concordance and ventriculoarterial discordance [1]. In D-TGA, the aorta arises from the morphologic right ventricle (RV), and the pulmonary artery (PA) originates from the morphologic left ventricle with parallel arterial trunks instead of the normal spiral association of the aorta and the PA [1,2]. The pathogenesis of D-TGA is due to a congenital conotruncal cushion defect during the embryonic period [3,4]. The reported incidence of D-TGA ranges from 20-30 per 100 000 live births and occurs predominately in males [5]. D-TGA is referred to as simple D-TGA in the absence of other related congenital anomalies [6]. D-TGA is categorized into 4 patterns: D-TGA with an intact ventricular septum, D-TGA with a ventricular septal defect (VSD), D-TGA with a VSD and an aortic arch obstruction, and D-TGA with a VSD and pulmonary valve stenosis (PS)/atresia [2]. The natural history of the disease has very poor outcomes, and probability to survive to adulthood without surgical repair is extremely low [7,8].

Surgical procedures have evolved from atrial switching (ie, the Mustard [9] or Senning procedures [10]) to arterial switching procedures (ie, Latané with or without the LeCompte maneuver) [11-13], and complex transpositions of the great arteries are often operated on using a Rastelli-type operation [14]. The Rastelli operation was first introduced in 1968 and soon became the standard surgical treatment for patients with a TGA, a VSD, and PS [14,15]. The VSD patch leads blood from the left ventricle to the aorta, and the RV is linked to the PA with a valved conduit. The 20-year survival rate has been reported to be <60% [8,16]. This report is of a 34-year-old Thai man who presented with pulmonary stenosis and heart failure 24 years after surgical correction with the Rastelli procedure for congenital D-TGA, VSD, and pulmonary atresia.

Case Report

A 34-year-old Thai man presented with dyspnea on moderate exertion for 2 months. Physical examination revealed mild tachypnea (ie, respiratory rate 24 breaths/min) and an oxygen saturation of 96% on room air. Finger clubbing was absent. His cardiovascular examination revealed a median sternal surgical scar, a parasternal heave, a grade III systolic ejection murmur at the left upper parasternal border, and a single second heart sound. No crepitations were superficially palpated at his chest wall. Mild pitting edema (ie, pitting edema 1+) at the pretibial area of both legs was detected. He had previously been diagnosed as having D-TGA with a VSD and pulmonary atresia and had undergone a modified right Blalock-Taussig shunt as a palliative shunt when he was 3 years old.

Figure 1. A 12-lead electrocardiogram shows sinus tachycardia, right axis deviation, deep S wave in V 5-6 leads compatible with right ventricular hypertrophy, and complete right bundle branch block.
At 9 years old, he had a brain abscess treated medically and surgically, including a right frontal craniotomy and a ventriculoperitoneal shunt. When he was 10 years old, he underwent a Rastelli-type operation with a 25-mm pulmonary homograft with a bifurcation PA plasty and double ligation of the right Blalock-Taussig shunt for definitive surgical correction. At that time, an intraoperative finding was that the aorta arose from the morphologic RV chamber, compatible with D-TGA, a large subaortic peri-membranous VSD, pulmonary atresia, and a confluent PA branch. After PA plasty, the main PA size was 18 mm, the right PA size was 11 mm, and the left PA size was 12 mm. After that, his condition was markedly improved. Thus, he did not visit the hospital regularly and was lost to follow-up because he could perform his activities of daily living without clinical dyspnea on exertion. His current medication before presenting to our hospital at 34 years old was only an antiepileptic agent that controlled his seizures well. During this latest visit, his electrocardiogram demonstrated sinus tachycardia, right axis deviation, and a deep S wave in the V6 lead, compatible with RV hypertrophy and complete right bundle branch block (Figure 1).

Transthoracic echocardiography and transesophageal echocardiography revealed degenerative calcification of the pulmonary valve (the RV-PA conduit), a pulmonary maximum velocity of 3.25 m/s with delayed peaking compatible with severe PS, marked RV hypertrophy and dilatation, mildly-impaired RV systolic function compatible with the long-standing pressure load, and tricuspid regurgitation with a maximum velocity of 5.1 m/s. There was no evidence of sub-valvular PS. Color M-mode Doppler flow propagation velocity showed turbulent flow across the interventricular septum from the left ventricle to the RV through the systolic phase, compatible with VSD patch leakage (Figure 2).

Figure 2. Transthoracic echocardiography demonstrates marked right ventricular dilatation (A), color Doppler M-mode in the parasternal long-axis view shows turbulent flow across the interventricular septum during the systolic phase compatible with ventricular septal defect patch leakage (B). Transesophageal echocardiography shows severe degenerative calcification with limited opening of the pulmonary valve (C) compatible with severe pulmonary stenosis (right ventricle-to-pulmonary artery conduit dysfunction) indicated by the yellow arrow.
A computed tomography (CT) scan of the heart demonstrated a calcified RV-PA conduit, an abrupt focal luminal narrowing at the origin of the bilateral main branch of the right PA (6 mm), and the left PA (11 mm), compatible with peripheral PS or supravalvular PS. The lesions of stenosis were demonstrated by the axial view and sagittal view of the CT scan (Figure 3). Three-dimensional reconstruction of the heart from the CT scan was useful in localizing the sites of stenoses involving the RV-PA conduit, located at the proximal ends of both the right pulmonary and left pulmonary arteries, and in understanding anatomical relationships between the adjacent structures (Figure 4). A right-left heart catheterization was performed to confirm the hemodynamic significance of pulmonary valve obstruction and branch PA stenosis. The right ventricular systolic pressure (RVSP) was 120 mmHg, the systolic main PA pressure was 106 mmHg, the systolic right PA pressure was 25 mmHg, and the systolic left PA pressure was 73 mmHg. The pullback gradient technique from both of the PA branches to the main PA and the main PA to RV confirmed the presence of significant valvular and supravalvular PS. Another finding was a significant step-up in mean oxygen saturation from the right atrium to RV level (63% to 71%, respectively). Therefore, either an intracardiac shunt at the ventricular level or VSD patch leakage with a left-to-right shunt was confirmed (Figure 5).

The final diagnosis in our patient was residual VSD patch leakage with valvular and supravalvular PS as long-term complications of D-TGA with VSD with pulmonary atresia after a Rastelli-type operation. After a multi-disciplinary team discussion with the patient, he opted for a third reoperation, and this was carefully planned to correct the long-term complications after the Rastelli-type procedure. The intraoperative findings for this reoperation were a severely-calcified pulmonary homograft as well as a bilateral proximal right PA stenosis and a left PA stenosis, both of which had severe calcification. VSD patch leakage at the RV outflow tract was also found. The patient underwent VSD closure and RV-to-PA reconstruction with polytetrafluoroethylene graft size 24 mm with PA plasty.
to correct the stenosis of both the right and left PAs. The patient was discharged 2 months after surgery. Echocardiographic evaluation showed improved RV function. At the time of writing, his status is partial recovery with mild limitation of physical activity (ie, a heart failure classification of New York Heart Association class II), and he is being followed up regularly at our cardiology out-patient clinic.

Discussion

The causes of right-sided heart failure include RV cardiomyopathies, RV ischemia, RV infarction, and increased ventricular volume loading due to valvular heart disease, and congenital heart disease and ventricular pressure loading due to PS or pulmonary hypertension due to a variety of reasons [17]. We highly suspected the present case would have PS due to his underlying disease of congenital heart abnormalities and abnormal results of the physical examination, including the systolic ejection murmur with a parasternal heave and the single second heart sound. Long-term complications after Rastelli-type operations in patients with D-TGA with VSD and pulmonary atresia include regurgitation or stenosis of the valved conduit between the RV and PA, residual VSD, and re-stenosis of the peripheral PA, which occurred in the present case. Other possible complications reported were an obstruction of the left ventricular outflow tract, aortic regurgitation, left ventricular dysfunction, arrhythmias, endocarditis, and sudden death [14,18-20].

Echocardiography is the initial diagnostic imaging modality that can visualize the connection between the junction of both of the ventricles and great vessels and assess the valvular function as well as the VSD. The structure and function of the conduit between the RV and pulmonary trunk must be evaluated with Doppler interrogation. The continuous wave from the Doppler measurements of the tricuspid regurgitation jet velocity is used to calculated RV pressure [20]. In many cases, echocardiography was of limited value in demonstrating the RV-PA conduit, the PA, and its proximal branches [21]. In such cases, additional imaging, such as cardiac CT or cardiac magnetic resonance imaging (cMRI), can be invaluable in demonstrating peripheral PA stenosis, as it was in the present case [22]. Invasive cardiac catheterization may be necessary for hemodynamic calculation of the conduit stenosis and to assess the level of stenosis [20]. An oxygen step-up of 5% between the right atrium and RV is diagnostic for a VSD with a left-to-right shunt. The differential diagnosis is patent ductus arteriosus with pulmonary regurgitation, ostium primum atrial septal defect, and coronary fistula to the RV [23]. However, there were no clinical clues suggestive of these in the present case.

Predictors of conduit failure from previous reports were a diagnosis of TGA, earlier age at RV-PA conduit implantation, and PA stenosis, all of which were present in our patient [24-27]. Previous reports state that around one-third of patients survived without needing a cardiac reoperation for conduit failure at 20-year follow-up [24,25].

Figure 4. (A, B) Three-dimensional reconstruction of the heart from computed tomography was useful in localizing the sites of stenosis involving the right ventricle-to-pulmonary artery conduit and in understanding anatomical relationships between adjacent structures. The sites of stenosis were the proximal ends of both the right pulmonary and left pulmonary arteries.
Figure 5. (A-C) The right-left heart catheterization in our patient demonstrated a high right ventricular systolic pressure of 120 mmHg compared with a left ventricular systolic pressure of 128 mmHg. The systolic pressure gradient across the right ventricle to the pulmonary artery (120 to 106 mmHg), the main pulmonary artery (106 mmHg) to both the proximal right pulmonary artery (25 mmHg) and the left pulmonary artery (73 mmHg) were also reported. These findings were compatible with significant right ventricular outflow tract obstruction and significant peripheral pulmonary stenosis. Another finding was significant oxygen step-up from right atrium to right ventricular level (63% to 71%, respectively). Therefore, either an intracardiac shunt at the ventricular level or VSD patch leakage with a left-to-right shunt was confirmed in our case.

Generally, reconstruction of the RV outflow tract in adults is performed using a homograft. However, satisfactory graft material is difficult to obtain and has the problem of degenerative calcification. Polytetrafluoroethylene conduits are a reasonable option for reconstruction of the RV outflow tract as a substitute for a pulmonary homograft [28,29].

A symptomatic patient with a RVSP > 60 mmHg is the indication for re-intervention of pulmonary valve conduits [20], and the present case had an RVSP of 120 mmHg, which was extremely high. In the current era of management, percutaneous pulmonary valve implantation has become the treatment strategy of choice for dysfunctional valves if it is technically achievable [20]. The challenge in the present case was that residual VSD
With supravalvular PS (peripheral pulmonary artery stenosis) correction was needed to treat the RV dysfunction. Thus, a third cardiac operation was the preferred choice in this situation.

After Rastelli-type operations, patients should have regular follow-up in a congenital heart disease center with expertise in adults at least once a year. The rate of event-free survival at 20-year follow-up was reported to be only 20%. Strenuous exercise should be avoided, but regular low-intensity exercise (eg, swimming, walking, and even cycling) should be encouraged [20]. Good dental hygiene should be encouraged to decrease the risk of infective endocarditis. Antibiotic prophylaxis is only suggested for high-risk patients, such as those with any prosthetic valve (including a transcatheter valve), those in whom any prosthetic material was used for cardiac valve repair, those who have had a previous episode of infectious endocarditis, those who are known to have any type of cyanotic congenital heart disease, and those who have had any type of congenital heart disease repair using a prosthetic material regardless of whether it was placed surgically or by percutaneous techniques for up to 6 months after the intervention or in their lifetime if there is a residual intracardiac shunt or residual valvular regurgitation [30].

**Conclusions**

We report the complications of and successful reoperation for a Rastelli-type operation at 24-year follow-up in a patient diagnosed as having D-TGA with VSD and pulmonary atresia. Long-term complications identified included VSD patch leak, RV-to-PA conduit stenosis, and branch PA re-stenosis. A multimodal imaging approach with the non-invasive and invasively investigations was useful in providing the most accurate preoperative diagnosis and in guiding the cardiac surgeon in planning for reoperation.

**Declaration of Figures’ Authenticity**

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