

D-Transposition of great arteries with severe calcific aortic stenosis in an adult patient: a rare presentation

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Kardiochirurgia i Torakochirurgia Polska 2022; 19 (1): 59-60

The incidence of transposition of great arteries (TGA) is 31 per 100000 live births worldwide [1]. It is, however, the most common cyanotic congenital heart defect seen in neonates [2].

The arterial switch operation (ASO) performed in neonates is the treatment of choice for patients born with TGA-intact ventricular septum (IVS). In India, however, late referral of patients with TGA and IVS is common. Left ventricle (LV) retraining followed by ASO is an option in late presenting TGA-IVS [3, 4]. Atrial switch as the primary procedure is another option.

We present the case of a 22-year-old man who was diagnosed with d-TGA-IVS and severe calcific aortic stenosis and was treated with a modified Senning procedure and aortic valve replacement. We believe this is the first case in the literature of d-TGA with right ventricular outflow tract obstruction (RVOTO) presenting this late. Institutional ethics committee approval and written informed consent from the patient were taken for data publication.

A 22-year-old man came with New York Heart Association (NYHA) grade II dyspnoea for 3 months. He had central cyanosis (SPO₂ 89%) and a grade 3/6 ejection systolic murmur in the aortic area. Interestingly, there was no history of any cardiac symptoms or workup.

Transthoracic echocardiography revealed d-TGA with a 34 mm ostium secundum atrial septal defect (ASD) resulting in a common atrium, bidirectional shunt, and ventriculo-arterial discordance. The aortic valve was thickened, sclerosed, and calcified with severe aortic stenosis with peak gradient/mean gradient (PG/MG) of 85/51 mm Hg. The aortic annulus was 24 mm. The aorta was to the right and anterior of the pulmonary artery. He did not have pulmonary hypertension and had normal biventricular function. The tricuspid valve regurgitation was more than mild.

Cardiac computed tomography (CT) showed the anomalous origin of the right coronary artery from the posterior sinus at the 6 o'clock position.

Pressures measured in cardiac catheterization were aortic 117/77(92) mm Hg, right ventricle (RV) 162/8(17), LV 47/6(16), left atrium (LA) mean (12), and pulmonary artery (PA) 40/15(23).

We performed a modified Senning procedure in which a baffle was created from the right atrium (RA) interatrial septum (IAS) junction to the floor of the LA using a polytetrafluoroethylene (PTFE) patch. The posterior lip of RA was anastomosed to the remaining LA to create a systemic venous baffle, so that venae cavae drained in the LV via the mitral valve. The pulmonary venous baffle was completed by attaching the anterior lip of the RA with the posterior lip of the LA using a pericardial patch so that pulmonary veins drained above the systemic venous chamber through the tricuspid valve to the RV. Aortic valve replacement was done with a bileaflet mechanical valve (St. Jude Medical 21 mm valve).

The patient had stable hemodynamics and no arrhythmias in the postoperative period and was discharged on day 7 on oral anticoagulants and antiplatelets. Post-operative echo showed a properly functioning baffle system with minimal gradients across the systemic venous baffle. The bileaflet mechanical aortic valve is also functioning properly, with a mean gradient of 18 mm Hg. The patient has been in regular follow-up since then and the six-month post-operative transthoracic echo showed similar findings as before. The patient is symptomatically better and maintaining SPO₂ 97%.

In d-TGA, the presence of a large ASD, ventricular septal defect (VSD), patent ductus arteriosus (PDA), or major aortopulmonary collateral arteries (MAPCAs) allows mixing of blood and survival beyond the neonatal period. The presence of a large ASD amounting to a common atrium allowed our patient to survive until 22 years of age. In our case, the patient's cyanosis went undetected most probably due to the dark complexion and unawareness of the parents and he became symptomatic only after the development of RVOTO (severe valvular aortic stenosis). On the other hand, d-TGA is associated with left ventricular outflow tract obstruction (LVOTO) more commonly.

In patients with d-TGA IVS presenting late, the most common problem is the failure of the systemic ventricle. These patients also develop significant atrioventricular valve insufficiencies. The management options for such patients are as follows: 1) two-stage or rapid two-stage ASO,

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Received: 5.08.2021, **accepted:** 23.01.2022.

2) direct ASO or Rastelli's procedure if there is associated pulmonary stenosis (PS) and if the LV is well developed, 3) atrial switch, 4) heart transplant or heart-lung transplant.

Late referral of patients with TGA-IVS is common in India. Only until the age of 9 to 10 years has LV retraining for executing ASO without prior atrial switch been documented in the literature [5]. In a case report for late presenting d-TGA IVS with right ventricular failure with pulmonary regurgitation (PR) in an 11-year-old patient, ASO with neo-aortic valve replacement was performed without LV retraining [6]. The option of primary ASO was thus not considered for our case as the presentation was extremely late. Similarly, the second option of direct ASO or Rastelli's procedure was ruled out due to the absence of an associated PS and the LV's loss of the ability to support the systemic circulation. A heart transplant was too far-fetched. Atrial switch as the primary procedure in the late presenting group is also an alternative to two-stage ASO with good results in long-term follow-up [7]. The oldest patient in a case series of modified Senning procedures performed for late-presenting d-TGA associated with total anomalous pulmonary venous connection (TAPVC) was 22 years old [8]. We, therefore, decided to go ahead with a modified Senning procedure for our patient.

Our case is unique because late presenting simple TGA with severe aortic stenosis (AS) has not been reported in the literature and was managed successfully with a modified Senning procedure and aortic valve replacement (AVR).

In conclusion, the modified Senning procedure is a good choice with limited risk for such very late presenting cases of d-TGA IVS. The management strategy must be tailored to

the individual. A long-term prognosis needs to be observed in such cases.

Disclosure

The authors report no conflict of interest.

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