

Atrial and visceral situs inversus with congenitally corrected transposition of the great arteries in a patient with dextrocardia, ventricular septal defect and pulmonary stenosis: A rare presentation

*Atriyal ve visseral situs inversusla birlikte doğuştan düzeltilmiş büyük damar transpozisyonu olan bir hastada dekstrocardi, ventriküler septal defekt ve pulmoner stenoz birlikteliği:
Nadir bir olgu*

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In this article, we present a 16-year-old female case of congenitally corrected transposition of the great arteries. The patient had atrial and visceral situs inversus with dextrocardia, and also ventricular septal defect with pulmonary stenosis. Presenting symptoms were cyanosis and dyspnea on exertion. The ventricular septal defect was closed through the left-sided morphological right atrium across the mitral valve. The surgeon operated from the left side of the patient. The pulmonary artery was augmented with a pericardial patch.

Key words: Atrial and visceral situs inversus; congenitally corrected transposition of great arteries; ventricular septal defect.

Congenitally corrected transposition of the great arteries with atrial situs inversus exhibiting dextrocardia is a rare anomaly.^[1,2] The ventricular outflow tracts in this anomaly do not cross, and the aorta and pulmonary artery lie parallel to one another. Dextrocardia is nearly always associated with this condition. There is fibrous continuity between the left-sided mitral valve and pulmonary valve, and the pulmonary valve is in a wedged position between the mitral and tricuspid valves. In congenitally corrected transposition of the great arteries with atrial situs inversus, the aorta is virtually always to the right. Ventricular septal defect is the defect most commonly associated with this condition. In addition, when atrial situs inversus is present, ventricular septal defect and/or subvalvular pulmonary stenosis are more

Bu makalede, 16 yaşındaki bir kız hastada nadir bir doğuştan düzeltilmiş büyük damar transpozisyonu olgusunu sunuyoruz. Hastada dekstrocardinin eşlik ettiği atriyal ve visseral situs inversus ve aynı zamanda pulmoner stenozlu ventriküler septal defekt vardı. Siyanöz ve egzersize bağlı dispne başlıca semptomlardı. Ventriküler septal defekt, mitral kapaktaki sol taraflı morfolojik sağ atriyum üzerinden kapatıldı. Cerrah, hastanın sol tarafından ameliyatı gerçekleştirdi. Pulmoner arter, perikardiyal yama ile büyütüldü.

Anahtar sözcükler: Atriyal ve visseral situs inversus; doğuştan büyük arterlerin düzeltilmiş transpozisyonu; ventriküler septal defekt.

common than with congenitally corrected transposition of the great arteries with atrial situs solitus.

CASE REPORT

A 16-year-old female patient presented to us with a history of cyanosis and dyspnea on exertion. A cardiovascular examination revealed a pansystolic murmur in the fifth intercostal space on the left side, and an ejection systolic murmur in the second intercostal space on the left side in the midclavicular line. Echocardiography and angiography (Figure 1-3) showed congenitally corrected transposition of the great arteries with atrial situs inversus. A chest X-ray revealed a gastric bubble on the right side suggesting visceral situs inversus. Operative intervention was planned.



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Figure 1. A cardiac catheterization study showing the catheter in the left-sided morphological right atrium.

On cardiopulmonary bypass with bicaval cannulation, the left-sided morphological right atrium was opened by a surgeon operating from the left side. There was a right superior vena cava in addition to a left-sided superior vena cava and an inferior vena cava, which was managed with a sump sucker in the coronary sinus. The ventricular septal defect was closed with a double velour

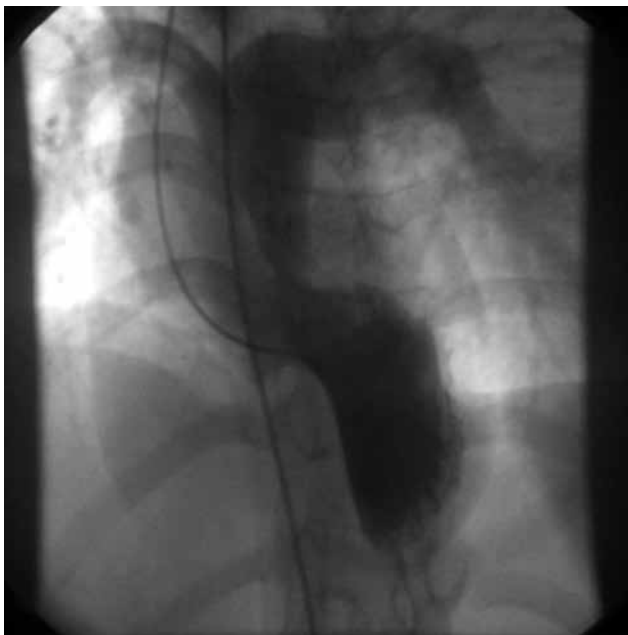


Figure 3. Ventriculography showing the morphological left ventricle ejecting into the pulmonary artery.

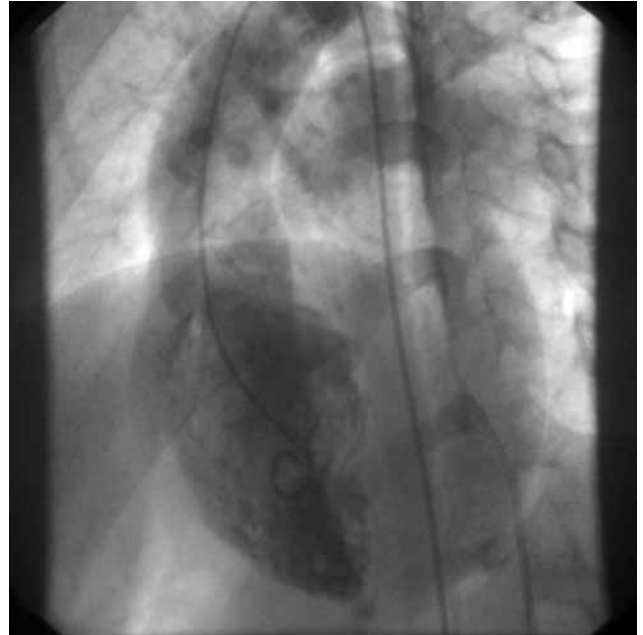


Figure 2. Ventriculography showing the morphological right ventricle ejecting into the aorta.

Dacron patch inserted through the right atrium across the mitral valve using interrupted pledgeted sutures. The pulmonary artery was opened, a commissurotomy was done, and a pericardial patch was used for transannular pulmonary artery augmentation. The patient came off the bypass with temporary pacing for a second-degree heart block. A permanent epicardial pacemaker was implanted on the 10th postoperative day for the second-degree heart block, and the patient was discharged two weeks after surgery.

DISCUSSION

The first repair of a cardiac anomaly associated with congenitally corrected transposition of the great arteries was performed in 1957 by Anderson, Lillehei, and Lester from the University of Minnesota.^[3]

Repairs of congenitally corrected transposition of the great arteries plus dextrocardia in adulthood^[4] have been reported in the literature. This case had atrial situs inversus with a bilateral superior vena cava along with congenitally corrected transposition of the great arteries.

Patients with congenitally corrected transposition of the great arteries in which the operation has been performed from the left side have been reported in the literature.^[5] In this case, because of the presence of atrial situs inversus and a small-sized right atrium, it was mandatory that the surgery be done from the left side.

Diagnosis is usually made via echocardiography, which can be confirmed by cardiac catheterization

or magnetic resonance imaging (MRI). In addition, MRI^[6] may be helpful in confirming other associated anomalies. A chest X-ray and ultrasonography of the abdomen are necessary to diagnose visceral situs.

Left ventricular outflow tract obstruction can be relieved with a transannular patch in patients with atrial situs inversus with congenitally corrected transposition of the great arteries since they usually have a normally positioned atrioventricular node^[7] and the bundle of His runs on the posterior inferior margin of the ventricular septal defect. This contrasts with the congenitally corrected transposition of the great arteries with atrial situs solitus in which the atrioventricular node is located anteriorly adjacent to the right atrioventricular valve orifice with the bundle of His coursing on the anterior right ventricular free wall caudal to the pulmonary annulus. It then descends to the anterior part of the infundibular septum.

Patients suffering from atrial situs inversus with congenitally corrected transposition of the great arteries are less prone to conduction anomalies compared with patients with atrial situs solitus with congenitally corrected transposition of the great arteries. However, in this case, the patient had conduction abnormalities postoperatively for which a permanent epicardial pacemaker was implanted.

Anatomical correction (double switch) was not possible in this case as there was severe pulmonary stenosis. The atrial switch using the Rastelli procedure was avoided because of problems associated with the prosthetic conduit^[8] and the complex nature of the repair.

In conclusion, for patients with atrial situs inversus congenitally corrected transposition of the great arteries with pulmonary stenosis, pulmonary artery augmentation can be done with a transannular patch, thus obviating the need for a prosthetic conduit. This is not possible for patients with atrial situs solitus congenitally corrected transposition of the great arteries with pulmonary stenosis.

The position of the chambers necessitated performing the operation from the left side.

Declaration of conflicting interests

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