Total Correction of Transposition of Great Arteries With Atrioventricular Septal Defect

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A successful anatomical correction of transposition of the great arteries with an atrioventricular septal defect is reported. This combination of anomalies is rare, and the anatomical correction is unusual in that the operation involves all four heart valves.


The association of atrioventricular septal defects with transposition of the great arteries (d-TGA) is rare [1-3]. These anomalies usually coexist with pulmonary stenosis or pulmonary atresia [3]. Unrestricted pulmonary blood flow is seldom seen in this complex of anomalies. We report a total anatomical correction in a patient with TGA and an atrioventricular septal defect (AVSD) with unrestricted pulmonary blood flow using the arterial switch procedure and a two-patch repair of the AVSD.

A 9-year-old girl was admitted with mild cyanosis apparently present from the age of 3 years. She was reasonably grown with height and weight on the 25th centile. Auscultation revealed a loud second heart sound with grade 3/6 systolic murmur and a soft diastolic grade 2/6 rumble in the third and fourth intercostal space of the left parasternal region.

Electrocardiogram revealed left-axis deviation with biventricular hypertrophy. Chest roentgenogram showed moderate cardiomegaly (cardiothoracic ratio, 26:36) with pulmonary plethora. Echocardiography and cardiac catheterization demonstrated situs solitus, atrioventricular concordance, and ventriculoarterial discordance with a well-developed confluent AVSD. There was a 16% increase in oxygen saturation in the right atrium with a further 8% increase in the right ventricle. The ventricular pressures were equalized (93/2 mm Hg), and the pulmonary artery pressure was at systemic level (84/55 mm Hg; mean, 65 mm Hg).

Operation was performed using standard cardiopulmonary bypass techniques and systemic hypothermia at 20°C. The aorta was anterior to and to the right of the pulmonary artery. The main pulmonary artery was dilated to three times the size of the aorta. The right coronary and the left anterior descending coronary arteries arose together from the left sinus, and a separate circumflex artery was found arising from the right sinus. There was an intermediate form of AVSD [3] with no cleft in the anterior leaflet of the mitral portion of the atrioventricular valve (Fig 1). Between the anterior portion of the mitral and tricuspid valves and the posterior leaflets of the mitral and tricuspid valves there was a layer of tissue on the summit of the ventricular septum with a well-developed ventricular septal defect.

Repair of the AVSD was done using the two-patch technique with two separate, fashioned bovine pericardial patches (Ionescu-Shiley, Irvine, CA) for closure of the ventricular and atrial components of the defect. The atrioventricular valve orifices were of adequate size and competence was achieved with minimal repair of the bridging leaflets at the point of attachment to the patch. The transposed great arteries were corrected by an arterial switch procedure with coronary transfer. The pulmonary artery was repositioned to the anterior root using the Le Compte maneuver. The coronary transfer was effected with a button of the aortic wall to the neoaoartc root. The resulting defect in the neopulmonary root was repaired with a pantaloon-shaped bovine pericardial patch. The discrepancy in the size of the distal aorta to the neoaoartc root was managed by adjusting the suture line and performing an anterior, vertical aortotomy to increase the sewing length of the distal aorta. It was possible to complete the anastomosis without use of prosthetic material.

The patient was weaned from cardiopulmonary bypass with minimal inotropic support. Prostaglandin infusion was commenced prophylactically to obviate elevation of pulmonary artery pressure. The pulmonary artery pressure fell to 75% of systemic level in the immediate postoperative period and dropped below 50% after extubation. The prostaglandin infusion was withdrawn without elevation of pulmonary artery pressure on the second postoperative day. An echocardiogram performed on the second postoperative day showed good biventricular function with mild regurgitation of the tricuspid valve. There was no residual shunt across the patch and no gradient across either the left or right ventricular outflow tracts. The patient, at 3 months' follow-up, is alive and well with no change in echocardiographic features.

Comment

The association of AVSD with TGA is rare. In a large series of 400 hearts with TGA there was only one with AVSD [1]. In a review of 132 cases of TGA, 2 had an AVSD.

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Fig 1. Schematic representation of transposition of the great arteries with atrioventricular canal. (a = posterior leaflet; Ao = aorta; ASD = atrial septal defect; b = anterior leaflet; c = right lateral leaflet; d = left lateral leaflet; LV = left ventricle; Pa = pulmonary artery; RA = right atrium; RV = right ventricle; VSD = ventricular septal defect.)

along with 1 case of AVSD, TGA, and patent ductus arteriosus (2.3%) [2]. In another large review of 507 specimens of hearts with AVSD, 17 were found to have complete TGA [3].

This anomaly is commonly reported with pulmonary atresia, or pulmonary valvular and subpulmonary stenosis [3]. In the same series the authors had reported only 1 case of an intermediate type where the anterior bridging leaflet was not free floating.

Surgical correction of this anomaly in hearts with pulmonary obstruction has been undertaken using a Rastelli [4] procedure or the Fontan principle [5]. Physiological correction using atrial redirection and AVSD repair has been carried out in 4 patients with AVSD associated with TGA by Pacifico and associates [6]. These authors repaired the defects using a modified Senning procedure in 2, modified Mustard operation in 1, and atrial baffle with ventricular inversion in 1. We performed total correction using both ventricles without a conduit.

The AVSD repair was carried out with two patches of bovine pericardium for closure of the defect without dividing the atrioventricular valve leaflets [7]. Only minimal suturing of the anterior and posterior leaflets of the mitral valve was necessary for its competence. The tricuspid valve was also similarly repaired. It is worth mentioning that the original pulmonary artery was three times the size of the aorta and considerable tailoring was needed for the anastomosis of great vessels to the new roots.

In view of the unrestricted pulmonary blood flow and systemic pulmonary artery pressure, the patient was prophylactically kept on prostaglandin infusion with continuous pulmonary artery pressure monitoring. Her pulmonary artery pressure stabilized at less than half systemic pressure with captopril therapy. As the patient did not have further elevation of pulmonary artery pressure, we believed that the pulmonary hypertension was reversible and would not be a problem in the long term. In this particular situation, the large ventricular component of the AVSD and the elevated pulmonary artery pressure almost certainly maintained left ventricular mass, which aided successful support of the systemic circulation after the arterial switch procedure.

We conclude that arterial switch with AVSD repair using both ventricles is feasible in anatomically favorable types of d-TGA with AVSD.

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References