BRIEF COMMUNICATIONS

SURGICAL REPAIR OF D-TRANSPOSITION WITH AORTOPULMONARY WINDOW: A CASE REPORT

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A 3.3 kg. white male neonate with dysmorphic features consistent with Goldenhar's syndrome had cyanosis at 8 hours of age. An echocardiogram revealed normal intracardiac anatomy with D-transposition of the great arteries (D-TGA), a type I¹ aortopulmonary window (APW), and a patent ductus arteriosus. The patient was begun on a regimen of prostaglandin E₁ infusion and underwent cardiac catheterization with balloon atrial septostomy on the second day after birth. Coronary artery anatomy was the type most commonly seen with D-TGA²: a two-vessel system with origin of the left vessel from the left cusp of the aortic (anterior semilunar) valve and of the right vessel from the posterior cusp of the aortic valve. The circumflex and anterior descending coronary arteries arose from the left main coronary artery. The origin of the left coronary artery was in close proximity and slightly inferior to the inferior margin of the APW (Fig. 1).

On the seventh day after birth, the patient underwent surgical repair under hypothermic (25° C) cardiopulmonary bypass. After ductal ligation and patch closure of the atrial septostomy, the great arteries were dissected free and the APW was located. The great arteries were transected at the level of the APW, with care taken to avoid injury to the aortic and pulmonic valves. The APW was transected at the same level. Right and left coronary ostia and surrounding buttons of aortic tissue were repositioned to the proximal portion of the neo-aorta after corresponding buttons of tissue had been removed. The pulmonary confluence was moved anterior to the aortic root and pulmonary continuity was reestablished. Defects in the neopulmonary artery from the excision of the coronary ostia were repaired with pericardial patches. The APW margins were incorporated into the suture lines during reanastomosis of the great arteries (Fig. 2).

The postoperative course was uneventful, with brief periods of junctional tachycardia (controlled with digoxin). Nitroglycerin and dobutamine were used for less than 24 hours. At his 12-month check-up, the patient was receiving no cardiovascular medications. An echocardiogram demonstrated normal ventricular size and function. There was mild

turbulence above the pulmonary anastomosis (estimated gradient 10 mm Hg) and no turbulence in the aorta.

Discussion. Patients with D-TGA and intact ventricular septum face a grim prognosis without surgical intervention.³ Use of Senning and Mustard intraatrial baffle techniques has resulted in acceptable surgical mortality rates but has left a legacy of right ventricular dysfunction and cardiac rhythm disorders. Improved initial mortality rates with the arterial switch procedure and better long-term results have established this procedure as the repair of choice for D-TGA. Associated structural pathology increases the complexity of and mortality associated with surgical repair; however, recognized structural contraindications are few, and the advantages of an anatomic repair are sufficiently great⁴ that arterial switch repair remains the method of repair for even complex D-TGA.

APW is an uncommon cardiac anomaly, representing less than 1% of cardiac lesions. APW coexisting with D-TGA is a rare association. Our review of the literature revealed only two other reported patients,^{5,6} neither of whom survived surgical repair. Tiraboschi and coworkers⁵ attempt⁵ to patch the APW (without addressing the D-TGA and ventricular septal defect) under conditions of bypass and hypothermia was further complicated by a second APW distal to the first. Krishnan and colleagues⁶

Fig. 1. Illustration of D-TGA of the great vessels, patent ductus arteriosus, and coronary artery relationship to the APW.

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Fig. 2. Arterial switch repair with the great artery transection at the level of the APW and incorporation of defect margins into the arterial reanastomosis.

reported patch closure of the APW and ventricular septal defect concomitant with a Senning atrial baffle repair. Both of these infants were first seen and were operated on at older ages (7 and 3 months respectively) than our patient, and both were found at necropsy to have significant pulmonary vascular disease. Accelerated progression of occlusive pulmonary vascular disease has been associated with d-TGA,⁷ particularly when combined with a left-to-right shunt.⁸ Coexisting D-TGA and APW should manifest a similar propensity toward early development of pulmonary vascular changes. Pulmonary hypertension has been suggested as a significant risk factor when assessing operability of APW⁹ and may be a significant factor in the outcome of operations performed on older patients.

The surgical repair described here provides the advan-

tages of a conventional arterial switch without extensive additional technical difficulty and is commonly performed before development of pulmonary vascular disease. Transection of the great arteries at the level of the APW in this case allowed the margins of the defect to be incorporated in the reanastomosis of the great vessels, and the coronary transfer could therefore be accomplished in the normal manner. A distal APW (Richardson type II or III) or an abnormality of coronary artery anatomy might not have been amenable to this surgical approach.

REFERENCES

1. Richardson JV, Doty DB, Rossi NP, Ehrenhaft JL. The spectrum of anomalies of aortopulmonary septation. *J THORAC CARDIOVASC SURG* 1979;78:21-7.
2. Elliott LP, Amplatz K, Edwards JE. Coronary artery patterns in transposition complexes. *Am J Cardiol* 1966;17:362-78.
3. Liebman J, Cullum L, Belloc NB. Natural history of transposition of the great arteries: anatomy and birth and death characteristics. *Circulation* 1969;40:237-62.
4. Van Praagh R, Jung WK. The arterial switch operation in transposition of the great arteries: anatomic indications and contraindications. *Thorac Cardiovasc Surg* 1991;39(Suppl):138-50.
5. Tiraboschi R, Salomone G, Crupi G, et al. Aortopulmonary window in the first year of life: report on 11 surgical cases. *Ann Thorac Surg* 1988;46:438-41.
6. Krishnan P, Airan B, Sambamurthy, Shrivastava S, Rajani M, Rao IM. Complete transposition of the great arteries with aortopulmonary window: surgical treatment and embryologic significance [Letter]. *J THORAC CARDIOVASC SURG* 1991;101:749-51.
7. Kumar A, Taylor GP, Sandor GG, Patterson MW. Pulmonary vascular disease in neonates with transposition of the great arteries and intact ventricular septum. *Br Heart J* 1993;69:442-5.
8. Newfeld EA, Paul MH, Muster AJ, Idriss FS. Pulmonary vascular disease in complete transposition of the great arteries: a study of 200 patients. *Am J Cardiol* 1974;34:75-82.
9. van Son JA, Puga FJ, Danielson GK, et al. Aortopulmonary window: factors associated with early and late success after surgical treatment. *Mayo Clin Proc* 1993;68:128-33.

ANTERIOR TRANSLOCATION OF THE RIGHT PULMONARY ARTERY TO PREVENT BRONCHOVASCULAR COMPRESSION IN A CASE OF TRUNCUS ARTERIOSUS AND TYPE A INTERRUPTED AORTIC ARCH

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The normal aortic arch produces a wide and harmonious curve that encircles the origin of the right pulmonary

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artery and the left pulmonary main bronchus. Relative shortening of the ascending or descending aorta (for instance after an arterial switch for transposition of the great arteries or after direct repair of a type A interrupted aortic arch) may produce a compression of these ele-

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