Surgical Closure of the Tricuspid Valve for Pulmonary Atresia, Intact Ventricular Septum, and Right Ventricle to Coronary Artery Communications

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SUMMARY. A surgical approach is reported for a patient with pulmonary atresia, intact ventricular septum, and right ventricle to coronary artery communications through sinusoids. A shunt procedure was performed at two days of age; the right ventricular outflow tract was not opened. At subsequent catheterization, the tricuspid valve was temporarily closed with a balloon catheter and no change was seen in the ECG. At five months of age, the right ventricle was plicated and a patch was sewn over the tricuspid valve. One year after surgery, neither the right ventricular cavity nor the sinusoids could be demonstrated at angiocardiography; ECG changes of left ventricular ischemia have resolved, and the child is growing normally.

KEY WORDS: Cyanotic congenital heart disease — Coronary “steal” syndrome — Pulmonary atresia — Coronary—camosel fistula

The optimal surgical approach to pulmonary atresia (PA) with intact ventricular septum (IVS) is controversial; when associated with right ventriculocoronary artery communications, the condition is often lethal. We report a case in which tricuspid valve closure was performed, converting PA with IVS into tricuspid atresia with intact ventricular septum.

Case Report

A female infant born at term weighing 3.2 kg had meconium aspiration and cyanotic heart disease. Cardiac catheterization at one day of age revealed pulmonary atresia, intact ventricular septum, tricuspid stenosis and regurgitation, hypoplastic right ventricle, and a patent ductus arteriosus. Right ventricular (RV) pressure was 82/13 and left ventricular (LV) pressure was 52/9. Right ventriculography (Fig. 1) filled the coronary arterial system through sinusoids. Balloon atrial septostomy was performed.

At two days of age, a 4-mm Impra tube graft was inserted side-to-side from the right subclavian artery to the right pulmonary artery. No attempt was made to open the right ventricular outflow tract.

A second cardiac catheterization was performed at four months of age. The diagnosis was unchanged and the shunt was functioning well. Right ventricular pressure was 125/14; left ventricular pressure was 88/7. The interatrial communication was large. With continuous external electrocardiographic monitoring, a no. 7 French end-hold balloon catheter was inflated in the right ventricle and pulled back to occlude the tricuspid valve. This procedure was continued for a total of 10 min without change in RV pressure, systemic pressure, or ECG. Dye injected into the right ventricle (Fig. 1) filled the left coronary artery and distal right coronary artery. The dye injection also confirmed that the tricuspid valve had been completely closed during the trial occlusion.

A second operation was performed at five months of age utilizing deep hypothermia and circulatory arrest. Patchy scar thinning of the epicardium was noted over the hypoplastic right ventricle. Through an oblique right atriotomy, pledgetted 4-0 Tycron mattress sutures were inserted to obliterate the right ventricular cavity. A Dacron patch 10 mm in diameter was sewn over the tricuspid valve. Postoperative course was smooth.

At 17 months of age, repeat cardiac catheterization showed continued wide patency of the shunt (Fig. 2) and an aortic saturation of 82%. Right atrial angiography did not opacify the right ventricle, and ascending aortography demonstrated normal coronary arteries without filling of sinusoids. Echocardiographic shortening fraction of the left ventricle was 30%; preoperatively this had been 27%. An improvement in the surface electrocardiogram was seen (Fig. 3) with resolution of the left precordial ST-segment and T-wave abnormalities noted before surgery.

Status of the right ventricular cavity was evaluated serially by echocardiography. Right ventricular end-diastolic volume, indexed to body surface area, was (cc/m²) 45.0 before surgery, 4.0 immediately after surgery, 7.4 at ten months postoperatively, and 5.7 at 18 months after surgery (2.9 cc/0.5 m²).

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Fig. 1. Pulmonary atresia, intact ventricular septum, and RV-coronary communications. A and B at catheterization at one day of age; C and D at 4 months of age. (A) Small RV cavity and hypoplastic regurgitant tricuspid valve are seen as well as sinusoids. (B) Early lateral view showing atretic RV outflow tract and anomalous coronary artery (white arrows) filling via sinusoids. (C and D) Anteroposterior and lateral frames with balloon (B) occlusion of the tricuspid valve. Black arrow shows atretic right ventricular outflow tract. Sinusoids are seen as well as distal right coronary artery and left coronary branches. RA, right atrium; RV, right ventricle; B, balloon.

Fig. 2. Angiograms after surgical closure of tricuspid valve. (A) “Great Ormond Street” type shunt is patent 17 months after insertion. (B) Contrast injection in the right atrium fails to opacify the right ventricle. (C) Ascending aortography demonstrates coronary artery branches but not the sinusoids.

Discussion

Pulmonary atresia with intact ventricular septum can be associated with sinusoidal communications between the right ventricular cavity and the coronary arterial system [1, 3]. These are considered persistent embryologic communications that do not involute because of intrauterine suprasystemic right ventricular pressure [7]. These abnormal communications deliver deoxygenated blood to the coronary arteries during systole and impair aortic diastolic filling of the coronaries because of delayed right ventricular ejection [2]. Pathologic effects of these communications include obstructive lesions within the coronary artery system (Fig. 3B [7]), endocardial sclerosis [2, 3, 7], and papillary muscle infarction [4]. Our patient also had patchy scarring of the epicardium noted at surgery.

Controversy exists over which of the following is optimal surgical therapy for the infant with PA with IVS: (a) shunt alone, (b) opening the RV outflow tract, or (c) combination of a and b. The decision-making process centers around the status of the RV and the tricuspid valve [5]. The presence of RV-coronary communications complicates the approach because excellent decompression of the right ventricle may create a coronary steal phenomenon (i.e., aorta to coronary artery to sinusoid to RV) [4, 6] and persistence of the hypertensive RV continues the coronary impairments discussed above.

Controversy is further complicated by lack of natural history data on the RV-coronary communications. Pathologic studies are preselected for deceased patients and important work by Patel et al. [5] deals only with survivors of operation. However, both Patel et al. [5] and Freedom and Harrington [2] have reported regression of the RV-coronary communications. While this appears more likely when RV decompression is achieved, two of six