Congenital tricuspid stenosis treated by a palliative open operation

Report of a case

The case is presented of a patient with congenital tricuspid stenosis who was treated successfully by a palliative open operation. The preoperative diagnosis was tricuspid atresia. At operation, however, congenital tricuspid stenosis was detected and managed by commissurotomy. The postoperative course was very good. Later, the associated ventricular septal defect (VSD), pulmonary annular stenosis, and probably the atrial septal defect (ASD) will be corrected. We emphasize the importance of diagnosing congenital tricuspid stenosis correctly, because of the possibility of operating upon the valve itself.

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Congenital tricuspid stenosis is a rare disease that is often incorrectly diagnosed, for it is easily confused with tricuspid atresia. Obtaining the correct diagnosis is important because the management for each case may differ.1 Only four cases of congenital tricuspid stenosis managed by surgery of the valve itself have appeared in the literature.1-5

The purpose of this paper is to present the case history of a patient with congenital tricuspid stenosis who was treated successfully by a palliative open operation. The case is of interest because it presented a specific pathology and required an unusual surgical approach.

Case report

E. F. S., a 5-year-old white boy, was admitted to the Heart Institute of the University of São Paulo Medical School on June 22, 1972. His symptoms included moderate cyanosis since birth and poor physical development. Dyspnea and hypoxia appeared when he was 6 months of age, becoming worse by the eighteenth month. Physical examination on admission showed poor somatic development, moderate cyanosis and clubbing of the fingers, a pulse rate of 100 beats per minute, and a blood pressure of 90/70 mm. Hg. A systolic murmur of medium intensity was audible over the left sternal border. There were no other symptoms worth noting.

The hematocrit value was 64 per cent and the hemoglobin, 21.8 Gm. per 100 ml. Chest roentgenography showed a slight decrease in the pulmonary circulation and moderate enlargement of the heart shadow along the expanse of the right atrium and great vessels. The left anterior oblique view showed "absence" of the right ventricle. The electrocardiogram revealed signs of left anterior hemiblock plus overloading of both atria and one ventricle; by vectorcardiography, we were able to determine that the left ventricle was the one involved.

Right heart catheterization was done (Table I), but it was impossible to pass the catheter into
the right ventricle. On cineangiography, the contrast media passed from the right atrium to the left atrium and ventricle, without opacifying the right ventricle (Fig. 1). Injection into the left ventricle showed a ventricular septal defect (VSD), slight hypoplasia of the right ventricle, and pulmonary stenosis (Fig. 2).

A diagnosis of tricuspid atresia Type Ia' was made, and the patient was operated upon on Sept. 15, 1972, via a median sternotomy. The right ventricle was moderately hypoplastic with tricuspid stenosis and an orifice 0.5 cm. in diameter (Fig. 3). Hypoplasia of the subvalvular apparatus and right ventricular inflow were also noted. An atrial septal defect (ASD) of 1 by 0.5 cm., a VSD of 2 cm., and a persistent left superior vena cava that drained into the coronary sinus were found. To correct the anomaly, we performed a tricuspid commissurotomy, enlarging the orifice to 1.8 cm. in diameter (Fig. 4), an infundibulectomy, and a pulmonary valvotomy. There were no immediate postoperative complications, and the patient was discharged in good condition.

The patient was asymptomatic and leading an almost normal life 8 months after the operation. Physical examination showed no cyanosis, disappearance of clubbing, and a decrease in the systolic murmur present in the pulmonary area. Blood tests revealed a hematocrit value of 47 per cent and a hemoglobin level of 15.7 Gm. per 100 ml. The electrocardiogram showed right bundle branch block, right ventricular overload, and no biatrial overload. The x-ray film showed a considerable increase in pulmonary circulation (as compared with preoperative flow) and an enlarged left ventricle. The left anterior oblique view still showed the image of absence of the right ventricle, although it was less noticeable.

Postoperative catheterization showed a bidirectional shunt at the atrial level (Table II) and systemic pressures in the right ventricle. A gradient pressure still existed between the right atrium and right ventricle. Cineangiography showed fair flow from the right atrium to the right ventricle and a pulmonary artery of good caliber (Fig. 5). The right ventricle is now of good size (Fig. 6).

Discussion

The definition of congenital tricuspid stenosis varies among authors. The term merely signifies fusion of the commissures for Medd and associates, who reserve the word hypoplasia for those cases in which narrowing of the ring is also present. However, for Riker and associates, congenital tricuspid stenosis implies narrowing of the ring plus shortening and thickening of the papillary muscles and chordae tendineae, in addition to fusion of the leaflets, which are usually thickened.

The incidence of congenital tricuspid stenosis is low. Abbott found only 3 cases in 1,000 autopsies of congenital heart disease. However, Svane found 5 in 394 such autopsies. Congenital tricuspid stenosis is always accompanied by a patent foramen ovale. However, one or more other heart malformations are often present and form the various complexes.

Hemodynamics

![Fig. 1. Preoperative cineangiogram. The catheter is introduced through the inferior vena cava and contrast injected into the right atrium. The enlarged right atrium, passage of contrast to the right ventricle and contrast injected into the right atrium.](image-url)
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The management for this anomaly is usually the same as for tricuspid atresia; that is, palliative treatment with one of the various systemic-pulmonary anastomoses. Even so, in some cases in which the right ventricle and pulmonary artery were of adequate size, corrective surgery was performed. For this reason, it is of great importance to distinguish between tricuspid stenosis and tricuspid atresia.

Medd and Kinmonth managed a case of congenital tricuspid stenosis and ASD by digital dilatation of the tricuspid valve and closure of the ASD. Riker and associates reported 1 case in which tricuspid commissurotomy and closure of the ASD were done and another in which transventricular pulmonary valvotomy and transatrial tricuspid dilatation were performed. Recently, Dimich and co-workers successfully treated a patient with congenital tricuspid stenosis and ASD by substituting a heterologous aortic valve for the tricuspid valve. Eight cases of isolated tricuspid stenosis have also been reported, but these lesions must have resulted from rheumatic disease of the tricuspid valve, even though the patients denied a history of rheumatic fever.

Our patient had marked tricuspid stenosis associated with an ASD, a VSD, and pulmonary valvular and infundibular stenosis. The right ventricle was moderately hypoplastic, so that total correction of the defects actually was inconceivable. Tricuspid commissurotomy and pulmonary valvotomy and infundibulectomy were therefore done. Some degree of pulmonary annular stenosis was intentionally left to protect the pul-

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**Fig. 2.** Preoperative cineangiogram. Contrast is injected into the left ventricle (LV). A large ventricular septal defect (VSD) and slightly hypoplastic right ventricle (RV) are seen. Ao, Aorta. PA, Pulmonary artery.

**Table II. Postoperative catheterization**

<table>
<thead>
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<th>Site</th>
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<th>D1</th>
<th>D2</th>
<th>MP</th>
<th>a</th>
<th>v</th>
<th>x</th>
<th>y</th>
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*Legend: Atrial waves are designated a, v, x, and y. For other abbreviations, see Table I.*
Fig. 3. Preoperative view of the orifice of the tricuspid valve (arrow) seen from the right atrium. The tip of a Kelly forceps introduced through the right ventriculotomy shows the size of the orifice.

Fig. 4. Postoperative appearance of the orifice of the tricuspid valve, which was enlarged by commissurotomy.

Fig. 5. Postoperative cineangiogram (right anterior oblique view). The catheter was introduced through the superior vena cava, right atrium, and tricuspid valve (TV) into the right ventricle (RV). Note the orifice of the tricuspid valve and filling of the aorta (Ao) and pulmonary artery (PA). Both the latter and the infundibulum have good caliber. LV, Left ventricle.
Postoperative cineangiogram (left anterior oblique view). Contrast was introduced into left ventricle (LV). Note the size of right ventricle (RV) on this film compared with the preoperative film (see Fig. 2). Ao, Aorta. LA, Left atrium. VSD, Ventricular septal defect.

The postoperative course was very good, and the patient was perfectly asymptomatic 8 months later. There are clinical signs of moderate pulmonary annular stenosis; however, the pulmonary artery appears to be of good caliber. A comparison of the preoperative and postoperative cineangiograms shows that the right ventricle is becoming larger (Fig. 6). We think this is an important point, because if the right ventricle expands to an adequate size, a truly total correction will later be possible. The increased mean atrial pressures (Table II) on postoperative catheterization could be explained by a moderate degree of myocardial depression. Thus, the final left ventricular diastolic pressure is higher whereas the systolic pressure is lower.

The patient will be followed periodically for later reoperation to correct the VSD and pulmonary stenosis.

The ASD should be closed only partially or not at all, because of the pressure gradient existing between the right atrium and the right ventricle due to the annular stenosis of the tricuspid valve.

The good result in the present case emphasizes the importance of the correctly diagnosing congenital tricuspid stenosis, operating upon the valve itself, and then correcting the associated defects in the first surgical approach. After development of the hypoplastic right ventricle, total correction in a second operation will be possible.

REFERENCES


