Congenital Cleft of the Anterior Tricuspid Leaflet With Severe Tricuspid Regurgitation in Adults

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Objectives and Background. Severe primary tricuspid regurgitation in the adult is a rare finding. This study describes the diagnostic findings and the treatment of an isolated congenital cleft of the anterior leaflet of the tricuspid valve as the morphologic substrate for severe tricuspid regurgitation.

Methods. The clinical, echocardiographic findings and the follow-up findings of five patients (all male, 20 to 56 years old) with this disorder are described. Four of the five patients underwent cardiac surgery that confirmed the diagnosis.

Results. In three of five patients, exertional fatigue was the limiting symptom (New York Heart Association functional classes II and III). The clinical findings included a holosystolic murmur and supraventricular arrhythmias in all patients. Cardiac catheterization, performed in four patients, yielded the incorrect diagnosis of Ebstein's anomaly in three. In one patient the cleft was associated with an atrial septal defect of the secundum type. In four of five patients successful reconstruction of the tricuspid valve with a DeVega annuloplasty was performed. One patient had a partial excision of the right atrium, and one had a closure of a coexisting atrial septal defect. One patient refused operation.

Conclusions. Tricuspid valve anomalies can be accurately identified by Doppler echocardiography. Surgical repair is the treatment of choice in patients with severe tricuspid regurgitation due to a congenital cleft of the anterior leaflet of the tricuspid valve.

(J Am Coll Cardiol 1992;20:1175-9)
axis view through the right side of the heart (Fig. 1). The diagnosis of an isolated cleft of the anterior leaflet implied correct insertion of the tricuspid leaflets. The presence of a defect in the anterior leaflet of the tricuspid valve was always associated with substantial regurgitant flow, as demonstrated by color-coded and continuous wave Doppler echocardiography.

**Results**

Five patients with a congenital cleft of the anterior leaflet of the tricuspid valve were recruited during a 7-year period with a total of 28,091 Doppler echocardiographic examinations. The Doppler echocardiographic diagnosis of this anomaly (Fig. 1) was confirmed during surgical exploration in four patients (Fig. 2).

**Clinical history.** In all five patients a heart murmur was first found between the ages of 12 and 56 years. At the time
of the first echocardiographic examination, the five male subjects (three Swiss, one Asian, one German) were 20 to 56 years old (Table 1). None of the patients had a history of endocarditis or severe chest trauma.

The cardiac history consisted of supraventricular arrhythmias in all patients (see laboratory findings or Table 1). Three of the five patients had signs of overt heart failure (New York Heart Association functional class II or III) (Table 1). Because of the severity of heart failure, one patient was admitted to our institution for evaluation of heart transplantation.

All patients had a 2–3/6 grade holosystolic murmur at auscultation. Three patients had a prominent V wave in the jugular venous pulse, and two had peripheral cyanosis. The physical examination revealed absence of other congenital abnormalities.

**Laboratory findings (Table 1).** All patients had arrhythmias. The electrocardiogram (ECG) documented chronic or paroxysmal atrial fibrillation in three patients, supraventricular premature beats in one patient and sinus tachycardia in another. In addition, three patients had complete and two had partial right bundle branch block. Two patients had first-degree atrioventricular block and one patient had a prolongation of the QT interval.

The physical exercise was tested by upright bicycle ergometry in four of five patients but was omitted in one patient because of manifest heart failure. In three patients working capacity was reduced by 50% to 71% of the predicted value and was normal in one patient (Table 1).

Chest X-ray film (Table 2) showed a marked enlargement of heart size in four of the five patients (Fig. 3) and was normal in one patient. A good echocardiographic image quality was obtained in all patients. Besides the cleft of the tricuspid valve, other cardiac abnormalities were found (Table 2): 1) One patient had an atrial septal defect of the secundum type, although the shunt was considered insignificant. 2) Similarly, in a second patient with a fistula from an atrial branch of the right coronary artery to the pulmonary artery, the shunt was not considered hemodynamically relevant. 3) A third patient who was not treated surgically had a dysplastic tricuspid valve; that is, the chordae tendineae of the septal and posterior leaflets were shortened and thickened. All patients had severe tricuspid regurgitation, but no pulmonary hypertension could be detected when calculated from the pressure difference between the right ventricle and right atrium with a continuous wave Doppler echocardiography.

Four patients underwent cardiac catheterization before the correct diagnosis was established by Doppler echocardiography; one patient underwent three catheterization studies. In three patients the catheterization findings were considered to indicate Ebstein's anomaly. Right heart catheterization and pressure measurements excluded pulmonary hypertension in the four patients studied (Table 2). Right ventricular angiography, performed in three patients, confirmed the presence of severe tricuspid regurgitation but failed to demonstrate cleft of the anterior leaflet as its underlying cause.

**Surgery and follow-up study.** Four patients had successful reconstruction of the tricuspid valve with DeVega annu-
loplasty (Fig. 2). In addition, partial excision of the right atrium was performed in one patient and closure of a coexisting atrial septal defect in another. Postoperative follow-up study was uneventful in three patients, and one patient is still being treated for heart failure (functional class II). The patient who refused operation remained asymptomatic with afterload-reducing medical treatment (functional class III).

Discussion

The clinical features of the described disorder include right heart failure, supraventricular arrhythmias and peripheral cyanosis. At cardiac auscultation, a grade 2-3/6 holosystolic murmur is audible. The ECG often demonstrates a right heart volume overload or right bundle branch block, or both, findings that are common in other forms of primary tricuspid regurgitation, such as dysplasias of the tricuspid valve (7). In most cases, the chest X-ray film shows marked enlargement of right-sided heart chambers without signs of pulmonary venous congestion. The invasive study confirms severe tricuspid regurgitation, whereas the cause of the regurgitation cannot be evaluated by conventional angiography. Echocardiography represents the most reliable diagnostic tool and allows description of the morphology of the tricuspid valve and a semiquantitative evaluation of the severity of the regurgitation. In particular, two-dimensional echocardiography accurately differentiates between Ebstein's anomaly, an endocardial cushion defect or other primary or secondary forms of tricuspid regurgitation. When transthoracic echocardiograms are of good quality, as they were in all of our patients, transesophageal echocardiography would provide no further information.

A cleft of the tricuspid valve is a rare Doppler echocardiographic finding. In a previous study (8) of adolescents and adults routinely examined with Doppler echocardiography, we found an incidence of one cleft in 161 patients with newly recognized congenital heart disease (0.6%). In the present study, five patients with this anomaly were found among 28,091 Doppler echocardiographic studies (0.018%). Even though all of our patients were male, because of the small size of the group, no conclusions relating to gender can be drawn.

Several efforts have been made to classify congenital tricuspid regurgitation. Lagarde et al. (9) separated patients with these anomalies into two groups. One group comprised the newborn, whose condition may be fatal within several days to weeks because of right ventricular failure (7) or may resolve without residual defects (10,11). In the latter case, the etiology is thought to be a functional overload of the right ventricle rather than a congenital anomaly (10,11). The other group comprised patients with decompensation during adult life, as demonstrated in our cases (8,9,12-15). In all cases the reason for the tricuspid regurgitations was dysplasias of the tricuspid valve.

Three of our cases were misdiagnosed as Ebstein's anomaly, which is the most common and best known cause of severe congenital tricuspid regurgitation. In Ebstein's anomaly, insertion of the septal and posterior tricuspid valve leaflets is dislocated into the right ventricle. This diagnostic feature is easily demonstrated by echocardiography.

Other, less common congenital disorders with isolated severe tricuspid regurgitation are dysplasias of the tricuspid valve. According to the classification of Becker et al. (16), the term dysplasias includes all deformities of the tricuspid valve that are not associated with an anomalous insertion of the tricuspid valve. They are divided into three subgroups (grades I to III), corresponding to the severity of the anomaly (Table 3). Thus, focal or diffuse thickening of the valve leaflets is classified as grade I, whereas a deficient development of the chordae and the papillary muscles or an improper separation of the valve components from the ventricular wall is summarized as grade II. A focal agenesis of valvular tissue is classified as grade III. Other rare forms of tricuspid malformations have been presented as case reports (17,18) or are described in our series. The cited classification (16) does not consider other anomalies of the tricuspid valve such as additional leaflets or clefts. The latter form of tricuspid valve anomaly represents a new morphologic entity and therefore deserves attention.

In contrast to clefts of the tricuspid valve, clefts of the mitral valve are well known. They are described as associated with atrial and ventricular septal defects, endocardial cushion defects, transposition of the great arteries and even as isolated anomalies (19,20). To date, no patient has been...
described with a congenital cleft of the tricuspid valve requiring surgical intervention for severe tricuspid regurgitation.

We hypothesize two possible explanations for the occurrence of a cleft. In all of our cases the cleft was located in the anterior leaflet close to the area where the lateral endocardial cushion meets the right dorsal conus swelling. The cleft might therefore be the result of an anomalous fusion. A second possible explanation is the large variability of the anatomic structure of the tricuspid valve itself. Under certain circumstances, even the pathologist finds it difficult to differentiate between clefts and additional valves (21).

A further speculation is based on the observation that one of the five patients with a cleft showed additional dysplasia of the tricuspid valve. With the assumption of an individual variation of one single malformation, this patient might have the full version of the anomaly, whereas the other four have a minor form of the dysplasia.

Conclusions. The newly described isolated cleft of the anterior leaflet of the tricuspid valve with severe tricuspid regurgitation is a rare congenital anomaly. It is probably a lone malformation of the tricuspid valve occurring during embryologic morphogenesis. The diagnosis is easily made by echocardiography. The treatment of choice is surgical reconstruction of the deficient leaflet.