CONGENITAL CLEFT OF THE ANTERIOR TRICUSPID LEAFLET WITH SEVERE TRICUSPID REGURGITATION IN AN ADOLESCENT

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Isolated congenital tricuspid valve incompetence is a very uncommon anomaly. This report describes the diagnostic findings and the treatment of an isolated congenital cleft of the anterior leaflet of the tricuspid valve. A 14-year-old male was operated upon with the diagnosis of atrial septal defect and regurgitation of the tricuspid valve. Atrial septal defect was closed with the primary suturing technique and successful reconstruction of the tricuspid valve with a DeVega annuloplasty was performed. Choice of the treatment is surgical reconstruction of the destroyed leaflet.

Key words: Tricuspid valve, cleft, anterior leaflet, atrial septal defect

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Severe primary tricuspid regurgitation in an adolescent is an unusual finding. Congenital tricuspid insufficiency due to a cleft in the anterior tricuspid leaflet is a rare congenital cardiac anomaly. Few cases are reported in the literature (1,2,3). However, most of the cases are associated with perimembranous ventricular septal defects, pulmonary stenosis or atrial septal defect (ASD). In this paper we reported a patient, who had been successfully operated for the cleft of the anterior leaflet which was associated with an atrial septal defect of the secundum type.

CASE REPORT

A 14-year-old male was referred to our institution for surgical repair. The patient was followed-up in an another hospital for ten years with the diagnosis of ASD. In the follow-up period, fatigue during exercise was the limiting symptom (New York Heart Association functional classes II). Cardiac manifestations were palpitations and early fatigue. The clinical findings included a holosystolic murmur on the mesocardiac region in the patient. The patient had undergone a basic investigation protocol which included chest X-ray, electrocardiogram, transthoracic echocardiography (TTE), bedside routine laboratory tests such as complete blood counts, creatinine, blood glucose, electrolytes, erythrocyte sedimentation rate. There weren't any electrocardiographic changes on the electrocardiographic recording. Chest roentgenogram showed non-specific findings. Laboratory tests were normal. Echocardiography was performed and showed an ASD (secundum type), pulmonary hypertension (45mmHg), regurgitation of tricuspid valve (2-3°), regurgitation of mitral valve (1-2°). Qp/Qs:1.9 was calculated (Figure I). Surgical correction was performed using bicaval and aortic cannulation, continuous cardiopulmonary bypass at 28°C, and crystalloid cardioplegia. Following right atriotomy, itwas seen that the cleft of the anterior leaflet was associated with an atrial septal defect of the secundum type (Figure 2). The tricuspid annulus was 35 mm in diameter.

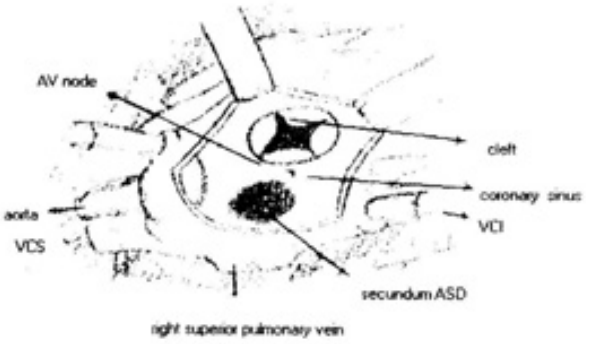

Figure 2. Schematic presentation of cardiac anomalies

There was no abnormality in the chordal attachment. The cleft was running through the anterior leaflet to the annulus of tricuspid valve. The cleft was repaired by three single bite using 6-0 polypropylene suture material. The ASD was closed with a primary suturing technique and successful reconstruction of the tricuspid valve was performed via DeVega annuloplasty (Figure 3). After rewarming and uneventful weaning from cardiopulmonary bypass, the operation was terminated in routine technique. The postoperative period was not complicated that the patient was extubated on 3trd postoperative hour and discharged from hospital on the 6th day without symptoms. Echocardiography before discharge demostrated grade 1 mitral incompetence and minimal incompetence of the tricuspid valve. Five months after the operation the patient's clinical status was excellent without any echocardiographic signs of tricuspid valve incompetence.

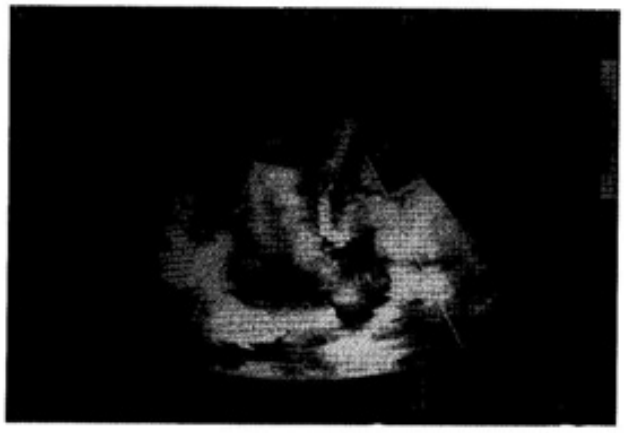

Figure 1. Preoperative echocardiography of the patient

Figure 3. I Schematic presentation of after reconstruction of the tricuspid valve and ASD

DISCUSSION

Diagnosis of tricuspid regurgitation can be established by two-dimensional echocardiography (4). Usually, tricuspid regurgitation can be described as primary, secondary or physiologic. Right ventricular volume or pressure overload can be the cause of secondary tricuspid regurgitation but tricuspid valve anatomy implies a normal condition. Physiologic tricuspid regurgitation is considered to be present in the absence of any primary or secondary form of tricuspid regurgitation (1). The causes of tricuspid regurgitation were described as follows; Ebstein' s anomaly, prolapse of tricuspid valve, rheumatic valve disease, endocarditis, trauma of the tricuspid valve, dysplasia of the tricuspid valve and dysfunction of tricuspid valve after right ventricular infarction (5). Isolated congenital anomalies of the tricuspid valve is a rare anomaly. Tricuspid regurgitation is a poorly discussed subject in the literature of cardiac surgery (1,5).

A cleft of the tricuspid valve is a rare Doppler echocardiographic finding (1). In a study of adolescents and adults routinely examined with Doppler echocardiography, an incidence of one cleft in 161 patients with newly recognized congenital heart disease (0.6%) was found (6). In another study, five patients with this anomaly were found among 28,091 Doppler echocardiographic studies (0.018%) (1). We couldn't describe the cleft of the tricuspid valve by Doppler echocardiography on this patient. The patient was operated with the diagnosis of ASD and tricuspid insufficiency. Several authors have classified the congenital tricuspid regurgitation. In a report, patients with these anomalies were divided into two groups (1). One group comprised of neonates, whose condition may be fatal within several days to weeks because of right ventricular failure or may resolve without residuel defects (1). In all cases the reason for the tricuspid regurgitation was the dysplasia of the tricuspid valve. Sometimes the case can be misdiagnosed as Ebstein's anomaly, which is the most common and best known cause of severe congenital tricuspid regurgitation. The diagnostic feature is easily demonstrated by echocardiography in Ebstein' s anomaly. In contrast to clefts of the tricuspid valve, clefts of the mitral valve are well known. They are known as associated with atrio-ventricular canal defects (1).

Eichhorn hypothesized two possible explanations for the occurrence of a cleft. In all of their 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 can be the result of an anomalous fusion. A second possible explanation is the large variability of the anatomic structure of the tricuspid valve itself (1).

Cleft of the anterior leaflet of the tricuspid valve is a rare congenital cardiac anomaly. The treatment is surgical reconstruction of the cleft.

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