Anatomic repair of Ebstein's anomaly with isolated anterior leaflet downward displacement

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Objective: Ebstein's anomaly with isolated anterior leaflet downward displacement is rare and has not been reported in the literature to our knowledge. In this article, our experience of the surgical treatment in 6 cases with this anomaly is reported.

Methods: From November 2005 to November 2013, 6 patients (3 male, 3 female, aged 2-39 years) with Ebstein's anomaly and isolated anterior leaflet downward displacement received anatomic repair at the First Hospital of Tsinghua University. The diagnosis was made by echocardiography and confirmed at operation. Surgery was performed under hypothermic cardiopulmonary bypass. Surgical technique included excision of a huge atrialized portion of the right ventricle located in the anterior wall of the heart; reconstruction of the right ventricle by repairing the "V"-shaped defect left by the excision procedure; detachment, repair, and reimplantation of the anterior leaflet; and reconstitution of the right atrioventricular connection. Intraoperative transesophageal echocardiography was used to evaluate the position, morphology, structure, and function of the tricuspid valve, as well as right ventricular function.

Results: Five patients were discharged uneventfully, and 1 patient died of postoperative pneumonia. At follow-up from 2 months to 7 years, no notable tricuspid valve regurgitation or stenosis was found and all patients were maintaining a normal lifestyle. The 27-year-old female patient gave birth to a normal infant uneventfully 3 years after surgery.

Conclusions: Ebstein's anomaly with isolated anterior leaflet downward displacement is a complex and severe abnormality, and has several unique anatomic and clinical features and specific surgical requirements. Preoperative diagnosis can be made by clinical investigation and echocardiography. Excellent results can be achieved by anatomic correction. (J Thorac Cardiovasc Surg 2014;148:1454-8)

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The most common morphologic change in Ebstein's anomaly (EA) is that the septal and posterior leaflets displace downward, with the anterior leaflet remaining in the normal position.[1,2]

It is rare for the anterior leaflet to displace downward in isolation with the septal and posterior leaflets remaining in the normal position, although both leaflets may be hypoplastic. There are no reports of cases with this kind of abnormality in the English or Chinese literature. The surgical techniques described for the more standard EA have not yet identified the morphologic abnormality found in this malformation. Six patients with this abnormality underwent anatomic repair at the First Hospital of Tsinghua University. Excellent results were achieved in 5 patients. Our experience with a novel surgical technique is reported.

CLINICAL DATA

From November 2005 to November 2013, 6 patients with EA and isolated anterior leaflet downward displacement (3 male, 3 female, age 2-39 years, weight 10-61 kg) received surgical treatment in the First Hospital of Tsinghua University. Among them, a 2-year-old boy was diagnosed by fetal echocardiography in the 38th week of pregnancy and then had poor development and low activity tolerance after birth. The other 5 patients presented with weakness, palpitations after activity, chest tightness, and frequent respiratory tract infections.

All patients were diagnosed through preoperative physical examination, electrocardiogram, chest x-ray, and ultrasonic echocardiogram (UCG). The electrocardiogram showed complete right bundle branch block in 3 patients, incomplete right bundle branch block in 2 patients, atrial fibrillation in 1 patient, and ventricular premature beat in 1 patient. Chest x-ray showed decreased pulmonary blood flow, right atrial enlargement, and cardiothoracic ratio of 0.58 to 0.90 (Figure 1, *A*). UCG was used to evaluate the morphology, position, structure, and regurgitation degree

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Abbreviations and Acronyms
ASD = atrial septal defect
CT = computed tomography
EA = Ebstein's anomaly
PFO = patent foramen ovale
RCA = right coronary artery
TV = tricuspid valve
UCG = ultrasonic echocardiogram

of the tricuspid valve (TV) (Figure 1, *B* and *C*), in addition to atrialized ventricle size, mitral valve function, and both right and left ventricular function. Three patients underwent computed tomography (CT) scan, which showed right ventricular enlargement, a giant atrialized right ventricle located in the anterior wall of the functional right ventricle, anterior leaflet downward displacement, and annulus enlargement. One patient underwent cardiac magnetic resonance imaging. The 29-year-old female patient was misdiagnosed with idiopathic right atrial dilatation at another institution by UCG and cardiac CT. One patient with this condition had an associated atrial septal defect (ASD), and 5 patients had a patent foramen ovale (PFO). Cardiac function of the 6 patients was New York Heart Association grade II to III. TV regurgitation was moderate in 2 patients and severe in 4 patients (Table 1).

METHODS

Anatomic surgical repair was performed in all patients. After sternotomy, the pericardium was opened, cardiopulmonary bypass was established routinely, and hypothermia was commenced. When the patient's core temperature reached 30°C, the aorta was clamped. Cold crystalloid cardioplegia solution was perfused via the aortic root for cardiac protection.

Intraoperative exploration confirmed that all patients had a huge atrialized ventricle located in the anterior of the heart with a thin wall forming a large cystic space (Figure 2). The inferior margin of the cyst abutted the anterior wall of the functional right ventricle at least 2 cm below the "V"-shaped defect. The anterior leaflet attached along the edge of the "V"-shaped defect was narrow, poorly developed, and significantly displaced downward. The posterior and septal leaflets were in a normal position in all patients. The TV annulus was enlarged and did not form a complete ring. It appeared that after repair, the papillary muscle positions and the size of the right ventricle would be almost normal.

After excision of the atrialized right ventricle, there was now an obvious "V"-shaped defect in the anterior wall of the right ventricle. The right coronary artery (RCA) ran along the inferior margin of the atrialized right ventricle in 5 patients and across the roof of the atrialized right ventricle in 1 patient. ASD was confirmed in 1 patient, and PFO was confirmed in 5 patients.

Excision of the redundant atrialized right ventricle wall, paying close attention to protection of the RCA and retention of part of the atrialized ventricular wall to establish a right atrioventricular connection was completed. As a result of excision of the giant atrialized right ventricle, the loss of integrity of the atrioventricular annulus and right ventricle caused by the huge "V"-shaped defect was revealed (Figure 3, *A1* and *A2*). It was necessary to suture the edge of the defect directly side-to-side to repair the right ventricular anterior wall and to construct a TV annulus of normal size to achieve normal geometry of the right ventricle. The anterior leaflet was detached from its base close to the defective edge of the anterior wall of the right ventricle. If the chordae tendineae were short and restricting the motion of leaflets, they were transected (Figure 3, *A1* and *A2*). To reconstruct the detached anterior leaflet, the detached base of the leaflet was enfolded by sutures to narrow this edge but lengthen the leaflet in its proximal-distal dimension. This resulted in an increase in the effective area of the leaflet when reattached in the normal annular position (Figure 3, *B* and *D*). A piece of fresh autologous pericardium was used to enlarge the poorly mobile septal leaflet in 5 patients (Figure 3, *C1* and *C2*). Care was taken to obtain proper coaptation of the 3 leaflets so that the TV exhibited no stenosis or insufficiency (Figure 3, *D*). Subsequently, the ASD or PFO was closed, suturing the remaining partially atrialized ventricle wall and right atrium wall to reestablish a stable right atrioventricular connection and achieve atrial closure to nearly normal size (Figure E1). The extended RCA was fixed to avoid kinking (Figure E2).

After evacuation of air from all chambers, the aortic clamp was released and the heart was restarted. There were no complications in weaning from cardiopulmonary bypass in any patient. The chest was closed, and surgery was completed (Table 2). The hemodynamic status of all patients remained stable in our group in the postoperative period with only a restricted use of dopamine and nitroglycerin. Central venous pressure was maintained at 6 to 8 mm Hg to minimize right ventricular preload.

RESULTS

Five patients recovered fully and were discharged uneventfully. One patient died of severe sepsis due to pneumonia 4 weeks after surgery.

Follow-up

The follow-up period was from 2 months to 7 years in 5 patients. UCG showed tricuspid leaflets in the normal position and functioning well with no stenosis in all patients, no regurgitation in 3 patients, and mild regurgitation in 2 patients (Figure 1, *D-F*). All patients were maintaining a normal lifestyle, and their cardiac function improved to New York Heart Association grade I. One patient gave birth uneventfully 3 years after surgery.

DISCUSSION

EA exhibits varied morphologic changes, which usually include atrialized right ventricle formation and tricuspid annulus enlargement. The leaflets may be hypoplastic and may have limited movement to different degrees in individual leaflets. It is most common that the septal and posterior leaflets are displaced downward, hypoplastic, or even absent, whereas the anterior leaflet is in a normal position. It is rare to find only the anterior leaflet abnormal and displaced downward while the septal and posterior leaflets remain in a normal position. To our knowledge, there is no report of this kind in the literature. In this group, there was a huge atrialized right ventricle to the anterior of the heart associated with several other unique pathologic features. Consequently, the surgical treatment must be different, even though the clinical symptoms may be almost the same as in most patients with EA.