Case Reports

Cone Reconstruction of Atypical Ebstein Anomaly

Associated with Right Ventricular Apical Hypoplasia

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Cone reconstruction for tricuspid valve repair has revolutionized the surgical treatment of Ebstein anomaly. We present the case of a 58-year-old woman with atypical Ebstein anomaly and right ventricular apical hypoplasia who was spared from palliative shunt physiology by our use of cone reconstruction.

Compared with other techniques, cone reconstruction of the tricuspid valve more closely replicates normal valvular anatomy and function. This surgical procedure can be applied to many anatomic variations of Ebstein anomaly, as in our patient's apparently unique instance of atypical Ebstein anomaly with right ventricular apical hypoplasia. ***(Tex Heart Inst J 2016;43(1):78-80)***

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Cone reconstruction (CR), or circumferential tricuspid valve (TV) repair, has revolutionized the surgical treatment of Ebstein anomaly.[1,2] We present the apparently unique case of a patient with atypical Ebstein anomaly and right ventricular (RV) apical hypoplasia who was spared palliative shunt surgery by our use of CR.

Case Report

In August 2013, a 58-year-old woman with a history of atrial arrhythmias and severe tricuspid regurgitation presented with fatigue and right-sided heart failure. Her medical history also included ulcerative colitis; superficial thrombophlebitis; and atrial flutter and atrioventricular nodal reentrant tachycardia, for which she had undergone successful radiofrequency ablation 3 years earlier. Results of a clinical examination included elevated jugular venous pressure, peripheral edema, and hepatomegaly.

The patient's treadmill exercise test results included average exercise capacity, normal heart-rate and blood-pressure responses to exercise, normal heart-rate recovery, no arterial desaturation, and a functional aerobic capacity of 101% of predicted value. A chest radiograph showed cardiomegaly. A transthoracic echocardiogram revealed reduced RV size and substantial hypoplasia of the RV apex (Fig. 1). The right atrium was markedly enlarged. Abnormal TV function included incomplete valvular coaptation, severe regurgitation, and annular dilation. Pulmonary valve cusp mobility was abnormal without regurgitation. The left ventricle (LV) was of normal size but occupied the apical portion of the heart, wrapping around the hypoplastic RV apex. The LV systolic function was normal; however, paradoxical septal motion was apparent.

Cardiac magnetic resonance images similarly revealed an enlarged right atrium, severe TV insufficiency, and RV apical hypoplasia (Fig. 2). The patient's LV ejection fraction was 0.59, and her RV ejection fraction was mildly depressed (0.46). Liver magnetic resonance elastograms revealed elevated stiffness at a value of 3.9 kPa, consistent with moderate hepatic fibrosis.

Preoperative cardiac catheterization was performed to delineate the patient's RV hemodynamic status and coronary anatomy. An RV angiogram showed RV apical hypoplasia with mildly to moderately decreased systolic function, severe tricuspid regurgitation, and a massively dilated right atrium (Fig. 3). On levophase, the patient's LV function was normal with no regional wall-motion abnormalities, and her coronary artery anatomy was normal. Atrialization of RV pressure waves was noted, and her RV end-diastolic pressure was elevated at 14 mmHg. Her pulmonary vascular resistance index was borderline elevated (4.7 WU*m^2), and her cardiac index was low (2 L/min/m^2).

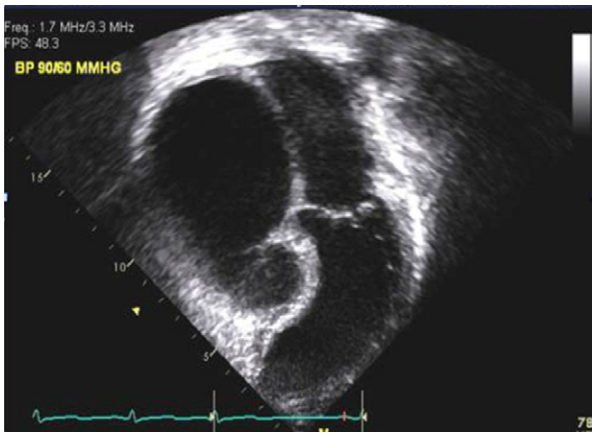

Fig. 1 *Transthoracic echocardiogram shows right ventricular apical hypoplasia. The left ventricular apex wraps anteriorly around the right ventricle. The septal leaflet displacement index appears to be normal.*

Supplemental motion image is available for Figure 1.

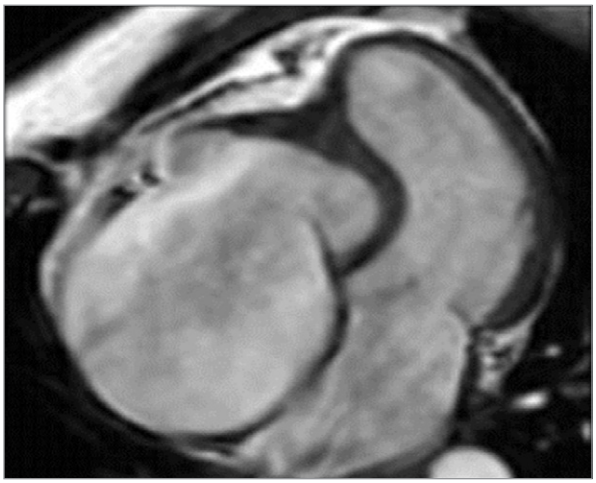

Fig. 2 *Magnetic resonance image shows severe tricuspid valve insufficiency, severe right ventricular apical hypoplasia, and a severely dilated right atrium.*

Supplemental motion image is available for Figure 2.

Intraoperative inspection of the TV yielded delamination of the septal leaflet. The anterior leaflet was tethered in multiple spots, and the inferior leaflet was diminutive. Overall, the anatomy was most consistent with Ebstein anomaly, given the areas of tethering between the anterior and inferior leaflets and the RV free wall; however, the septal leaflet tissue was not substantially displaced.

We performed TV repair by means of circumferential CR: 360° leaflet-tissue repair anchored at the true TV annulus after anterior-leaflet delamination or detachment from the RV free wall. Additional procedures included augmentation of the anterior leaflet with use of a porcine membrane patch, placement of a 28-mm flexible annuloplasty ring, and right reduction atrioplasty. A postoperative echocardiogram revealed trivial tricuspid regurgitation without stenosis. Ten months postoperatively, the patient's peripheral edema had improved, consequent to diuretic therapy. As of April 2015, an echocardiogram showed mild tricuspid regurgitation without stenosis; on clinical examination, the patient had residual hepatomegaly and mildly improved jugular venous pressure.

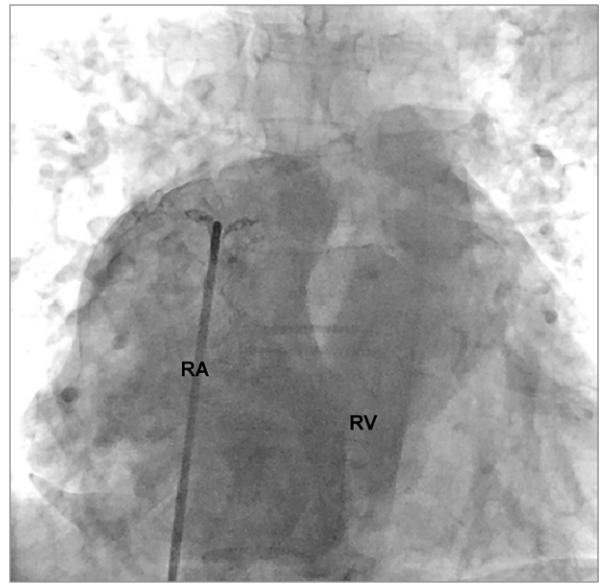

Fig. 3 *Right ventricular angiogram (anteroposterior view) shows a severely dilated right atrium (RA), severe tricuspid regurgitation, and an apically hypoplastic right ventricle (RV).*

Supplemental motion image is available for Figure 3.

Discussion

In Ebstein anomaly, the TV leaflets adhere to the myocardium because of failed delamination. Typically, the anterior portion of the TV annulus is positioned more inferiorly, is not displaced, and is often redundant or sail-like.[2,3] The result is a division of the RV into a functional RV and an atrialized RV, the result of downward displacement of the functional tricuspid annulus.[2,3] This anatomic anomaly results in incomplete coaptation of the TV leaflets and consequent tricuspid regurgitation. Over time, this leads to progressive enlargement of the right atrium and the atrialized RV.[2,3]

To our knowledge, ours is the first report of Ebstein anomaly with RV apical hypoplasia. In our patient's atypical form of Ebstein anomaly, some expected findings of the condition were present, including a diminutive inferior leaflet and tethering of the anterior leaflet. However, the septal leaflet was not tethered or displaced and the RV was not dilated, as would usually be seen; in fact, the RV was very small, with apical hypoplasia. The

authors of the only other report that we found of atypical Ebstein anomaly described the patient's anatomy as a dilated right atrium with isolated apical displacement of the inferior TV leaflet and normal insertion of the septal and anterior leaflets.[4] In several case reports of isolated RV hypoplasia, no author described Ebstein-like features.[5-7] Most of the patients who had isolated RV hypoplasia underwent palliative surgery that involved systemic-to-pulmonary artery shunts, bidirectional cavopulmonary anastomosis, or total cavopulmonary (Fontan) connection. Our patient was spared surgical palliation by virtue of definitive repair involving CR.

Cone reconstruction has revolutionized the treatment of Ebstein anomaly. Data from our institution have shown promising early results after CR.[1] The procedure, which involves reconstruction of the TV at the true anatomic annulus, imparts valvular function that is more like normal valvular anatomy than the function imparted by any previously described technique.[1] The CR technique can be applied to many anatomic variations of Ebstein anomaly, as in this instance of atypical Ebstein anomaly with RV apical hypoplasia.

We think that CR should be considered as a primary treatment when appropriate, instead of the default palliative procedure. However, certain clinical situations, such as severely depressed RV function, might warrant palliation.

References

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