Case of Ebstein Anomaly Complicated by Left Ventricular Outflow Tract Obstruction Secondary to Deformed Basal Septum Attributable to Atrialized Right Ventricle

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A 59-year-old woman presented to Okinawa Chubu Hospital with a several-month history of progressive dyspnea. The patient initially noted exertional dyspnea after walking only a half-mile or when lifting heavy objects. Dyspnea worsened progressively, resulting in multiple syncopal episodes. The patient denied chest pain or dyspnea at rest. Her past history was significant for Ebstein anomaly, hypertension, hyperlipidemia, and duodenal ulcer. Her current medication included furosemide 20 mg twice a day, spironolactone 25 mg every day, losartan 25 mg every day, and pravastatin 10 mg every day.

On initial examination, her blood pressure was 100/75 mm Hg, her heart rate was 75 bpm, and her respiratory rate was 18 breaths/min. A grade 4 systolic ejection murmur was noted throughout the precordium. Jugular venous pressure was not elevated and peripheral edema was not present. A chest roentgenogram showed cardiomegaly (Figure 1A) without pulmonary vascular congestion. An ECG showed normal sinus rhythm and marked left ventricular hypertrophy as manifested by high voltage and strain-type ST-T changes in I, aVL, V6 through V4 (Figure 1B). Laboratory tests showed normal kidney and liver functions and no evidence of anemia, and brain natriuretic peptide was 399 pg/mL (<18.4 pg/mL). An echocardiogram showed the apically displaced septal leaflet of the tricuspid valve in comparison with the anterior leaflet (Figure 2A and 2B), consistent with Ebstein anomaly. Functional right ventricle was so diminutive, and there was moderate tricuspid regurgitation by color Doppler. Aortic and mitral valve morphology appeared normal. Basal septum above the attachment of the septal leaflet of the tricuspid valve (atrialized right ventricle) was significantly deformed, resulting in abnormal protrusion of the basal septum toward the left ventricular outflow tract (LVOT). Systolic anterior motion of the anterior mitral leaflet was noted. Doppler evaluation revealed a high-pressure gradient at LVOT (Figure 3A through 3D, Movie I in the online-only Data Supplement). Severe mitral regurgitation was also noted (Movie II in the online-only Data Supplement). No atrial or ventricular septal defect was identified. These anatomic abnormalities were confirmed by MRI (Figure 2C and 2D).

Cardiac catheterization confirmed the hemodynamic abnormality (Figure 4). The pressure gradient at LVOT was as high as 130 mm Hg, whereas there was no pressure gradient across the aortic valve. After a premature ventricular contraction, LVOT gradient went up to >200 mm Hg and aortic pressure dropped to 80 mm Hg. Pulmonary artery wedge pressure, and pressures in the pulmonary artery, the right ventricle, and the right atrium, as well, were normal. Cardiac output was decreased at 2.12 L·min⁻¹·m⁻². The left ventricular ejection fraction was 64% and there was moderate to severe mitral regurgitation. The coronary arteries were normal. No evidence of left-to-right shunt was noted at the atrial and ventricular level. The final diagnosis was Ebstein anomaly complicated with severe LVOT obstruction secondary to protrusion of the basal septum as the result of the deformity of the atrialized right ventricle.

Discussion

Ebstein anomaly is a rare congenital anomaly of the tricuspid valve and the right ventricle occurring in 1 per 200,000 live births. This anomaly is characterized by (1) adherence of the septal and posterior leaflets to the underlying myocardium, (2) downward displacement of the functional annulus (septa I>posterior>anterior), and (3) dilatation of the atrialized portion of the right ventricle. Associated cardiac malformations include atrial and ventricular septal defect, pulmonary atresia or hypoplastic pulmonary artery, Wolf-Parkinson-White syndrome, etc. In addition, abnormalities of the left side of the heart have been known to develop in Ebstein anomaly, including noncompaction of the left ventricle, accessory mitral valve tissue, abnormal muscle bands, endomyocardial fibroelastosis, and mitral valve prolapse. LVOT obstruction associated with Ebstein anomaly is very rare. In a review article from the Mayo Clinic, Attenhofer Jost et al reported that 2 of 106 patients with Ebstein anomaly had myectomy for hypertrophic obstructive cardiomyopathy. Edwards from the same institution reported an autopsy case of severe Ebstein anomaly in which the left ventricle was compressed by the dilated atrialized right ventricle, resulting in the leftward...
bowing of the ventricular septum. In addition, Isobe et al reported an adult case with LVOT obstruction secondary to abnormal accessory tissue of the mitral valve. The present case may be similar to the hypertrophic cardiomyopathic state reported from the Mayo Clinic. Our case had a definite (>20 mm) downward displacement of the septal leaflet of the tricuspid valve, resulting in a large atrialized RV that protruded or bowed toward the LVOT. This deformity of the septum, in turn, significantly contributed to the development of the severe pressure gradient across LVOT, mimicking hypertrophic obstructive cardiomyopathy and mitral valve regurgitation secondary to systolic anterior motion of the anterior mitral leaflet.

Figure 1. A, A chest roentgenogram showing marked cardiomegaly. B, A 12-lead ECG showing marked left ventricular hypertrophy.

Disclosures
None.

References
Figure 2. A, A parasternal long-axis view indicating marked deformity of the basal septum protruding toward LVOT as shown by the larger arrow. A smaller arrow indicates the attachment of the septal leaflet of the tricuspid valve. B, An apical 4-chamber view showing the same finding as in A. C, A contrast-enhanced MRI showing marked downward (apical) displacement of the attachment of the septal leaflet of the tricuspid valve (smaller white arrow), whereas the attachment of anterior leaflet is normal (black arrow). The enlarged atrialized right ventricle resulted in the protrusion of the septum toward LVOT (larger white arrow). D, A contrast-enhanced MRI at a slightly higher level of the heart. The findings are the same as in C. LVOT indicates left ventricular outflow tract.
Figure 3. A through C, Apical 5-chamber views with (B and C) and without (A) color Doppler indicating that the obstruction of LVOT (A and B) and mitral regurgitation (C) were caused by the protruded septum and systolic anterior motion of the anterior mitral leaflet (an arrow in A), resulting in acceleration of the flow velocity up to 5 m/s (a white arrow in D, indicating that LOVT gradient was as high as 100 mm Hg gradient). A white arrowhead indicates a Doppler signal secondary to mitral regurgitation. Ao indicates ascending aorta; ARV, atrIALIZED right ventricle; LA, left atrium; LV, left ventricle; and LVOT, left ventricular outflow tract.
Figure 4. Pressures obtained at the time of cardiac catheterization. The left ventricular pressure was as high as 250 mm Hg and there was 130 mm Hg of LVOT gradient, whereas wedge pressure was not elevated. After a premature ventricular contraction, the LVOT gradient increased up to 300 mm Hg, whereas the aortic pressure dropped below 100 mm Hg with spike-dome configuration (aortic pressure was obtained from the femoral sheath). There was no pressure gradient across the aortic valve. There was no pressure gradient in the ascending aorta and femoral artery, either. Ao indicates aorta; LV, left ventricle; LVOT, left ventricular outflow tract; and WP, pulmonary artery wedge pressure.
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Circulation. 2016;133:e33-e37
doi: 10.1161/CIRCULATIONAHA.115.016208
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/133/4/e33

Data Supplement (unedited) at:
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