strictive pericarditis, systolic ventricular function is usually well maintained, despite impaired compliance, so that echocardiogram demonstrates normal or increased functional indices and a small left ventricle with a small cardiac output. This was clearly seen in our patient; good excursion of the septum and posterior ventricular wall resulted in good systolic functional indices, although small left ventricular dimensions owing to reduced ventricular compliance resulted in small stroke volume.

In summary, extreme respiratory variation in the depth of the pulmonic "a" wave was demonstrated in a patient with subacute constrictive pericarditis. This echocardiographic feature correlates with the respiratory response of mild constriction. Echocardiographic disparity in systolic and diastolic ventricular function is characteristic of constriction and important in ruling out restrictive cardiomyopathy, in which both systolic and diastolic ventricular function are concomitantly impaired.

**REFERENCES**

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**Partial Atrioventricular Canal in Association with Ebstein’s Anomaly**

**Echocardiographic Diagnosis and Surgical Correction**

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Echocardiography was used to diagnose the presence of both Ebstein’s anomaly and partial atrioventricular canal in the case of a 20-year-old man who presented for evaluation of supraventricular tachyarrhythmias. The diagnosis was confirmed at surgery with successful surgical repair.

**Ebstein’s anomaly and atrioventricular canal malformation** is a rare combination of congenital cardiac lesions. Kilby et al. reported such a case in a 13-year-old girl who died on the day following repair by the “atrial well technique” in 1956. The diagnosis was made at surgery, and no autopsy was performed. Lev et al. have described one pathologic specimen with tetralogy of Fallot, complete atrioventricular canal and a tricuspid valve “reminiscent of Ebstein’s complex.” No case of isolated atrioventricular canal defect and Ebstein’s anomaly diagnosed under cardiopulmonary bypass or by pathologic examination has been reported.

Reported is the combination of Ebstein’s anomaly and partial atrioventricular canal, ie, ostium primum atrial septal defect with a left anterior mitral valve leaflet which was diagnosed by wide angle, two-dimensional echocardiography and found at operation. Although cardiac catheterization revealed partial atrioventricular canal, this procedure did not confirm the presence of Ebstein’s anomaly.

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CASE REPORT

A 20-year-old man with documented episodes of supraventricular tachycardia since age 6 years was admitted for evaluation of palpitations and dizziness.

Physical examination revealed a slender white man in no acute distress with a pulse rate of 70 beats/min and a blood pressure of 110/68 mm Hg. The jugular venous pressure was not elevated, and the contour revealed “A” wave dominance. The carotid pulse was normal. There was a prominent chest wall deformity with bulging in the left precordial area. A prominent parasternal right ventricular lift was present and separate from the left ventricular impulse located just lateral to the midclavicular line. The second heart sound remained split on expiration. There were S₃ and S₄ gallops. There was a grade 3/6 harsh systolic murmur, heard best approximately 2 cm lateral to the left sternal border, which increased with inspiration and during the Müller maneuver. There was a grade 3/6 higher pitched holosystolic murmur heard best at the apex, which increased with isometric handgrip. There was a grade 1-2/6 middiastolic rumble not initiated by an opening snap, heard best over the lower left sternal border.

The ECG demonstrated Wolff-Parkinson-White abnormality. Two-dimensional echocardiogram revealed right-sided heart enlargement together with Ebstein’s anomaly and an ostium primum atrial septal defect (Fig 1). A different recording angle with benefit of a movable M-mode cursor from the two-dimensional echocardiographic machine revealed a marked delay in the closure of the tricuspid valve (Fig 2). The small cleft in the mitral valve, later seen at catheterization, was not visualized by echocardiography. The chest x-ray film demonstrated right ventricular enlargement with increased pulmonary vascularity consistent with a left-to-right shunt.

Cardiac catheterization revealed mildly elevated pressures in the right side of the heart with normal pulmonary arteriolar resistance and normal left ventricular pressures. Oxygen saturation determinations revealed a left-to-right shunt at the atrial level providing a pulmonary-to-systemic blood flow ratio of 3.4:1. The left ventriculogram demonstrated 2+ mitral regurgitation, with abnormal mitral valve position in the left ventricular cavity causing a “goose-neck” deformity of the left ventricular outflow tract.

Figure 1. Two images obtained from apical four-chamber position with corresponding diagrams. Left panel best demonstrates the dilated right atrium (RA) and right ventricle (RV) with displacement of tricuspid valve into right ventricle. Anterior leaflet of tricuspid valve (ATV) is markedly elongated with multiple ectopic chordal attachments to free wall of the right ventricle. Septal leaflet of tricuspid valve (STV) originates from the right surface of the ventricular septum (VS) and is displaced interiorly approximately 15 mm within right ventricle. Anterior leaflet of mitral valve (AL) originates from tip of ventricular septum; no ventricular septal defect could be visualized. Right panel demonstrates a normal secundum portion of atrial septum together with a large ostium primum atrial septal defect (ASD). LA, indicates left atrium; LV, left ventricle; I, inferior; S, superior; R, right; L, left; and PV, pulmonary veins.

Figure 2. This M-mode recording appears to show tricuspid valve (TV) (top arrow) closing approximately 110 msec after mitral valve (MV) closure (bottom arrow). This could be obtained only through ventricles while orienting sector beam through inflow tract of right ventricle and in a short axis tomographic plane of left ventricle.
anterior leaflet of the mitral valve had a cleft deformity. A ventricular septal defect could not be demonstrated. The right ventriculogram revealed tricuspid regurgitation. The tricuspid valve motion and attachments could not be adequately assessed.

At surgery, externally the heart showed no apparent atrialized portion of the right ventricle, although there was right atrial and right ventricular enlargement and hypertrophy. However, upon opening the right atrium, the tricuspid valve was observed to be grossly abnormal. The large tricuspid anulus was filled with disorganized sheets of valvular tissue with two separate orifices. The anterior valvular tissue clearly inserted distal to the A-V groove (Fig 3). There was an outflow primum atrial septal defect and a cleft anterior mitral valve leaflet. There was also a patent foramen ovale. After closure of the cleft mitral leaflet and repair of the ostium primum atrial septal defect with a pericardial patch, cardiopulmonary bypass was discontinued without difficulty. The patient exhibited normal sinus rhythm without any degree of heart block. Because of the minimal tricuspid regurgitation exhibited preoperatively, nothing was done to the tricuspid valve. Postoperatively, the patient’s course was uneventful, and he was discharged on the eight postoperative day. He has been physically active at home without symptoms or recurrent arrhythmias. Follow-up two-dimensional contrast echo study reveals no evidence of any residual shunting at the atrial level. The mitral valve appears unchanged, with normal motion.

**DISCUSSION**

This case is unusual, not only because of the rare association of Ebstein’s anomaly along with partial form of atroventricular canal, but also because the diagnosis was made by two-dimensional echocardiography. The two-dimensional study demonstrated abnormal attachments of the septal-appearing tricuspid leaflet, along with an elongated, redundant anterior leaflet which had multiple attachments to the right ventricular free wall by ectopic abnormal chordae tendineae (Fig 1). This pattern has been previously described to be indicative of Ebstein’s anomaly of the tricuspid valve.6

Conventional M-mode echocardiographic visualization of tricuspid and mitral valve leaflets did not adequately visualize the tricuspid valve, however, with counterclockwise rotation and slight medial-inferior angling of the transducer, a recording was made which demonstrated the markedly delayed closure of the tricuspid valve (Fig 2). The tricuspid valve appeared to close approximately 110 m sec after mitral valve closure; such a delay ≥40 m sec is usually considered indicative of Ebstein’s anomaly.4,5 At the time of surgery, the diagnosis of Ebstein’s anomaly in association with partial atroventricular canal was documented. Although the cleft in the mitral valve could not be seen by two-dimensional echocardiography, this was a very small defect as documented at surgery.

In conclusion, we have presented a case of partial atroventricular canal defect in combination with Ebstein’s anomaly in whom the diagnosis was made by two-dimensional echocardiography and confirmed at surgery. Two-dimensional echocardiography is the most accurate means of making this diagnosis preoperatively.

**REFERENCES**