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CASE REPORT

Abiomed Biventricular Assist Device as a Bridge to Transplantation in a 14-Year-Old With Cardiomyopathy Resulting from Ventricular Noncompaction

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Abstract Ventricular noncompaction is a rare but well-documented cause of cardiomyopathy. This report presents a case of ventricular noncompaction diagnosed late in end-stage cardiac failure and malignant ventricular arrhythmia, which required an Abiomed biventricular assist device as a bridge to transplantation.

Keywords Cardiomyopathy · Orthotopic heart transplantation · Ventricular assist device · Ventricular tachycardia

Ventricular noncompaction is a rare form of cardiomyopathy, currently categorized as an unclassified cardiomyopathy by the World Health Organization. This disorder often occurs in adults, with only a few series reporting pediatric cases. We present a delayed presentation of ventricular noncompaction in a teenager, with rapid deterioration of cardiac reserve necessitating emergent support using a biventricular assist device (BIVAD) and ultimate transplantation.

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Case Report

A 14-year-old previously healthy boy presented to an outside hospital emergency room with shortness of breath of 2 days duration. He had experienced progressive exercise intolerance and worsening shortness of breath over a 2-month period, which had been attributed to reactive airway disease. A chest radiograph showed cardiomegaly, and he was transferred to our institution for further workup and management.

At arrival, he was found to have ventricular bigeminy, and an echocardiogram showed a markedly dilated left ventricle with an end-diastolic diameter of 70 mm, global hypokinesis, a shortening fraction of 15%, and echocardiographic evidence of ventricular noncompaction. Very prominent trabeculations were noted in the inferior apical left ventricular posterior free wall. The ratio of the noncompacted to compacted myocardium during systole measured approximately 2 to 1, consistent with noncompaction.

He was started on a milrinone infusion, and a right heart catheterization was performed, which showed a cardiac index of 1.9 l/min/m^2 and a right atrial pressure of 5. His pulmonary artery pressures were 1/2 systemic, and his pulmonary arterial wedge pressure was 16 and his pulmonary vascular resistance was calculated at 3 Wood units.

He was intubated. An intraaortic balloon pump and a Swan-Ganz catheter were placed. Anti-failure therapy was maximized, and he was listed for transplantation. However, he experienced runs of ventricular tachycardia with significant hemodynamic compromise, which necessitated emergent implantation of an Abiomed extracorporeal biventricular assist device (BIVAD) (Abiomed Inc., Danvers, MA, USA).

He was rapidly weaned off his inotropic and pressor support and extubated in 3 days. He had excellent

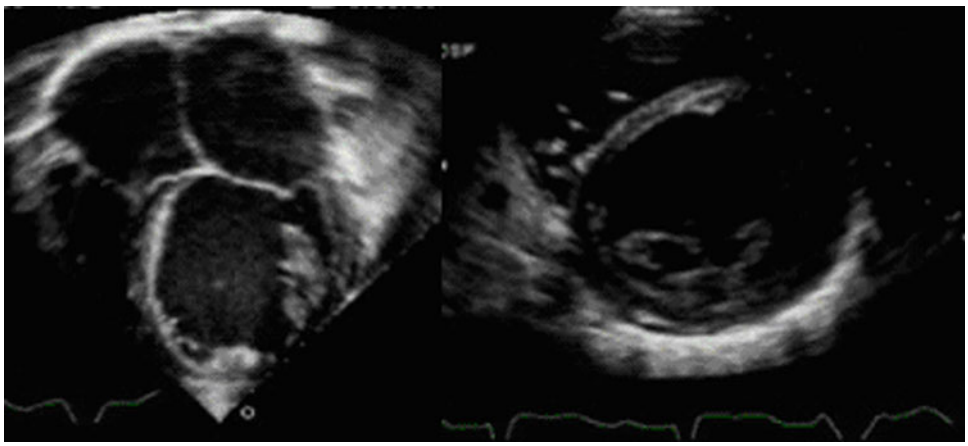

Fig. 1 Echocardiographic features of left ventricular noncompaction in four-chamber and parasternal short-axis views

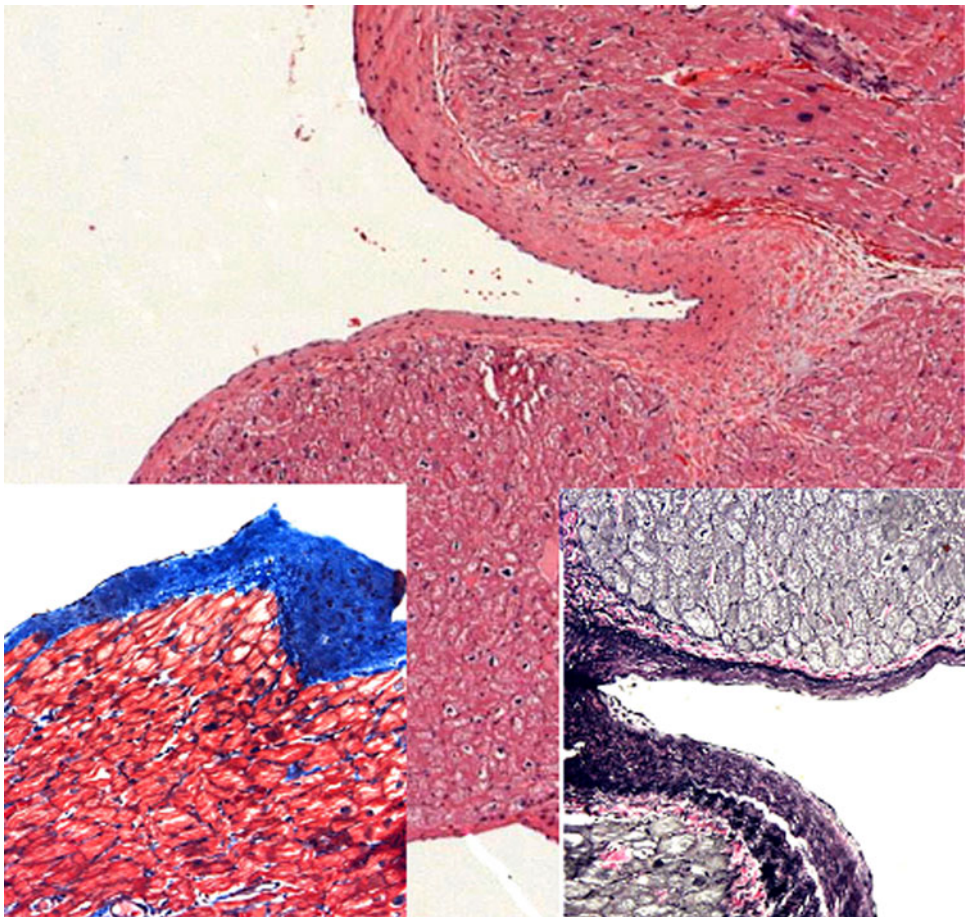

Fig. 2 Histologic appearance of the spongiform left ventricular myocardium with thickened endocardium on hematoxylin and eosin staining and on trichrome (*blue insert*) and elastin (*purple insert*) staining

end-organ function recovery after device placement, with normalization of renal function. He was successfully transplanted within 5 days of the BIVAD implantation, with excellent postoperative hemodyamnics and end-organ function. His immediate postoperative period was complicated by recurrent right lower lobe collapse and a right pneumothorax, which resolved with aggressive chest physical therapy and placement of a thoracostomy

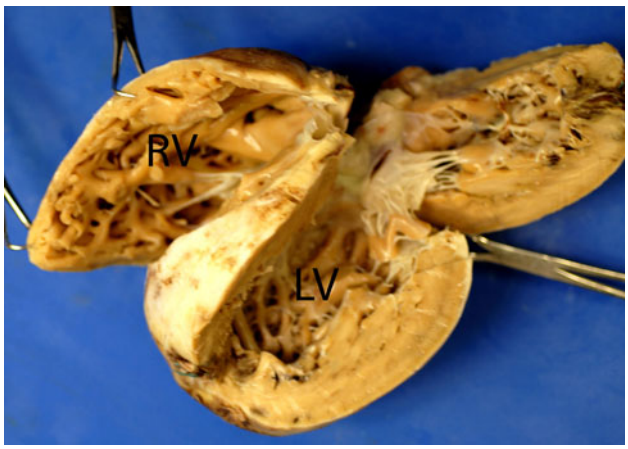

Fig. 3 Macroscopic appearance of the explanted heart clearly outlining hypertrabeculation of the left ventricular myocardium

tube, respectively. He was discharged home 30 days later and has done well. His most recent echocardiogram showed good biventricular function.

Discussion

Ventricular noncompaction is a rare cause of dilated cardiomyopathy. It occurs due to an intrauterine arrest of the normal myocardial compaction process [1, 2, 5, 6, 12]. The incidence of ventricular noncompaction is reported to be approximately 1.26% [5], with two-thirds of patients presenting with congestive heart failure, one-fifth presenting with arrhythmias, and about one-sixth presenting with thromboembolic events [5]. The reported mortality rate is 7 to 14% [5, 8].

The etiology of congestive heart failure in ventricular noncompaction is multifactorial. Noncompacted myocardium is more dependent on aerobic metabolism and thus more sensitive to hypoxia. Noncompaction also can cause abnormalities in the coronary microcirculation, with resultant subendocardial ischemia and fibrosis [4, 5, 12]. Echocardiography usually is diagnostic, although computerized tomographic angiograms (CTA), magnetic resonance imaging (MRI), and magnetic resonance imaging angiography (MRA) are gaining favor [1, 3, 7, 8, 10–12].

Both sporadic and familial forms of ventricular noncompaction exist [1]. A few reports have described orthotopic heart transplantation for this disease [8, 9]. In their pediatric series, Pignatelli et al. [8] reported an orthotopic heart transplantation rate of 11%. To date, no report has described the use of a biventricular assist system in the management of this rare entity (Figs. 1, 2, 3).

Conclusion

Ventricular noncompaction is a relatively rare cause of dilated cardiomyopathy in the pediatric population. The reported case illustrates the indolent course this disease may take, resulting in delayed diagnosis and the need for urgent mechanical support of the failing heart.

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