Noncompacted Ventricular Myocardium: Is Syncope the Only Warning Sign?

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Abstract: We present a case of a 14-year-old young boy with ventricular noncompaction and coexistent Wolff-Parkinson-White syndrome who presented with syncope. Detailed testing was performed, and an ablation was successful for Wolff-Parkinson-White syndrome; however, because of patient/family desires, an implantable cardiodefibrillator was placed. A careful review of the literature highlights the complex decision making involved with this condition.

Key Indexing Terms: Wolff-Parkinson-White syndrome; Isolated noncompaction of the ventricular myocardium; Cardiomyopathy. **[Am J Med Sci 2010;339(5):497–500.]**

Although the incidence of isolated noncompacted ventricular myocardium remains unknown, awareness of this entity has become increasingly widespread, largely because of improved imaging and identification of this cardiomyopathy. Noncompaction of the ventricular myocardium is characterized by prominent trabeculation of the left ventricular (LV) (and potentially the right myocardium) thought to result from incomplete embryogenesis. Thus, there can be associated congenital cardiac defects and neuromuscular disorders.[1,2] Clinically, these patients are primarily at risk for heart failure, arrhythmias, and serious thromboembolic events. In fact, electrocardiographic abnormalities are common including Wolff-Parkinson-White syndrome (WPW).[3–5] The incidence of WPW syndrome varies from different reports, but the incidence of WPW syndrome (15%) is higher in children with noncompaction.[6] The incidence of sudden death is not known but certainly increased compared with that expected for a matched population.

Herein, we report a young patient with ventricular noncompaction and WPW syndrome who presented with syncope. A careful review of the literature illustrates some of the difficult decision making required for this newly described condition.[2]

CASE REPORTS

A 14-year-old African American boy, a previously healthy adolescent, was admitted to Memorial Hermann Hospital with the first episode of syncope that occurred while running. The patient did not have any chest pain, palpitations, dyspnea, lightheadedness, or dizziness before his syncope. He had no family history of sudden cardiac death (SCD) or syncope nor was there any early heart disease in primary family members. He was on no medications and had no significant medical history. On presentation, his pulse was 70 beats per minute (bpm), blood pressure was 108/70 mm Hg, and his respirations were 16/min. He was afebrile; he was not orthostatic. He was alert and oriented with some superficial abrasions over his left forehead and left arm. His cardiac examination was unremarkable. His baseline laboratory examination was normal. His baseline electrocardiogram (ECG) is shown in Figure 1.

His ECG revealed sinus rhythm with heart rate of 66 bpm, with a PR interval of 80 milliseconds, and an apparent delta wave consistent with an accessory pathway diagnostic of WPW syndrome. Based on surface ECG, the accessory pathway is characterized as a posterior pathway. The 24-hour Holter monitor indicated only sinus rhythm, with rates of 53 to 134 bpm with a mean rate of 75 bpm. The heart rate responses to reported activity were appropriate. Normal diurnal variations in heart rates were observed. Preexcitation was consistently present with short PR interval and a permanent delta wave. A 2D echocardiogram revealed normal LV function with multiple thickened LV trabeculations consistent with ventricular noncompaction. Subsequent magnetic resonance imaging provided further detail demonstrating LV hypertrophy and numerous pronounced trabeculations (Figure 2). There was no evidence of LV thrombus. Because of the patient's presentation with syncope, an electrophysiologic study was performed with successful ablation of the accessory pathway.

At baseline, the rhythm was sinus with preexcitation, and the characteristics of the pathway were considered marginal risk, and no supraventricular or ventricular tachycardia was induced. Accessory pathway could not be demonstrated at the completion of the study. The postablation ECG is shown in Figure 3. An extensive discussion with the patient, his family, and all involved providers was then undertaken to describe the outcomes of patients with ventricular noncompaction and how it applies to this exact case. Ultimately, the decision to proceed with an implantable cardiodefibrillator (ICD) was agreed on and successfully placed via the left subclavian vein. The patient recovered uneventfully and has been placed on long-term anticoagulation. He has been asymptomatic since his initial admission.

DISCUSSION

This case highlights many of the complex decisions that may be encountered when a patient is diagnosed with ventricular noncompaction. Although isolated LV noncompaction was first described in 1984,[7] recent increased imaging and awareness has resulted in this entity being recognized by a host of medical societies as a genetic cardiomyopathy.[8,9] However, ventricular noncompaction remains as an unclassified cardiomyopathy according to the World Health Organization.[10] More importantly, ventricular noncompaction has become an important entity to consider when diagnosing and managing cardiomyopathy. Ventricular noncompaction is in a broad population of patients characterized by excessively prominent trabeculations and deep intertrabecular recesses in the left ventricle without other congenital cardiac abnormalities.[3] In normal

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Submitted January 20, 2010; accepted in revised form February 10, 2010.

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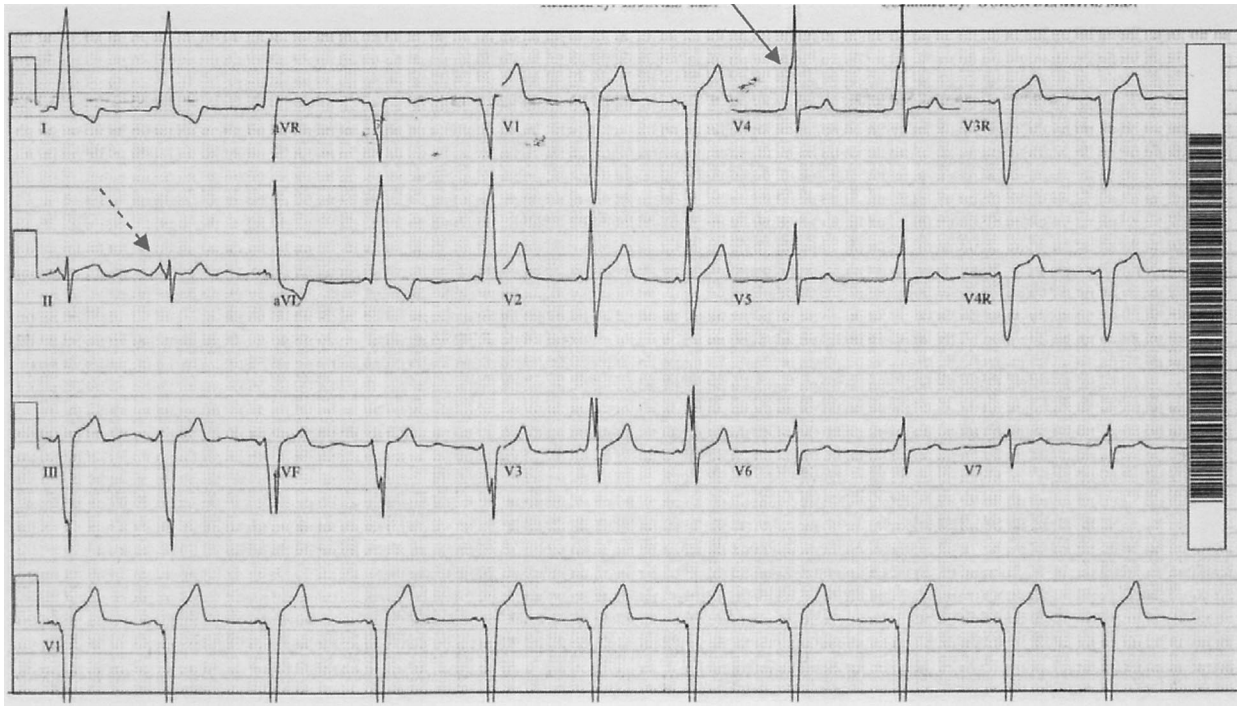

FIGURE 1. ECG at baseline. Solid arrow shows delta wave. Dashed arrow shows short PR interval. ECG, electrocardiogram.

developmental process, the direct continuity between the atrial and ventricular myocardium is lost when the annulus fibrosus forms.[11] The basic morphogenic abnormalities of ventricular noncompaction are thought to be caused by the arrest of the normal process of compaction and normal process of development of the annulus fibrosus.[3] Interestingly, the defects in annulus fibrosus may allow the formation of the accessory pathways.[12] As a result, ventricular noncompaction has been associated with multiple abnormal ECG patterns typically associated with accessory pathways such as WPW, left bundle branch block, left axis deviation, and other arrhythmias including atrial fibrillation.[3–5] In fact, the most frequent clinical manifestations of symptomatic patients with ventricular noncompaction are ventricular arrhythmias, heart failure, and systemic embolizations,[3,4,13–15] which accounts for excess mortality and morbidity associated with this condition. Thromboembolic events

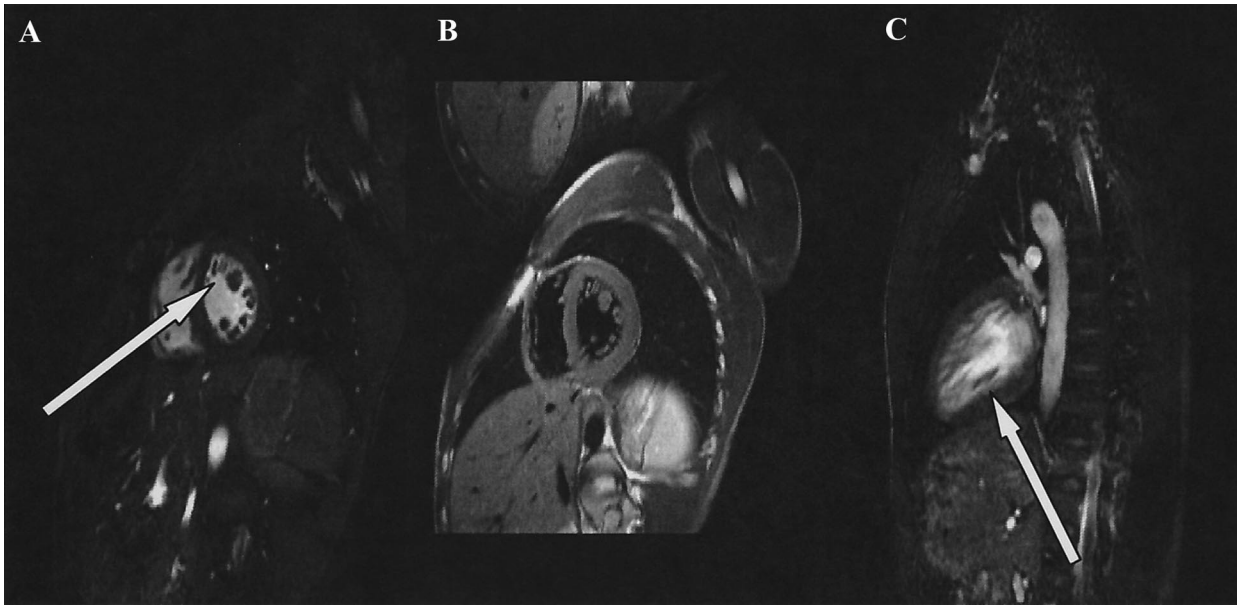

FIGURE 2. Magnetic resonance imaging (MRI) in a patient with ventricular noncompaction. Short axis (A), longitudinal (B) images, and sagittal MRI images (C) showing no thrombus but increased trabeculation of the left ventricle (white arrows).