Cardiology in the Young

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A boy with fatigue and heart block: what's the mechanism?

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Brief Report

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Abstract

When the atrioventricular node is damaged, accessory pathways can perform primary atrioventricular conduction but may spontaneously degrade during childhood. After surgical atrial septal defect repair during infancy, an adolescent male presented with fatigue due to iatrogenic complete atrioventricular node block with a degrading antegrade accessory pathway resulting in symptomatic bradyarrhythmia.

Case presentation

An 11-year-old boy was evaluated for fatigue and decreased exercise tolerance over the six months prior to presentation. At nine months of age, at an outside institution, he underwent surgical repair of an ostium primum atrial septal defect using a single pericardial patch. At that time, he was noted to have a cleft mitral valve without evidence of ventricular septal defect. Although ostium primum atrial septal defects are commonly associated with left axis deviation, the patient was reported to have a normal electrocardiogram preoperatively. The postoperative electrocardiogram reportedly revealed a new left bundle branch block QRS pattern. A routine electrocardiogram completed two years prior to our initial evaluation was described as unchanged from the prior electrocardiograms since the procedure (Fig. 1). However, it demonstrates a short PR interval, delta wave, and a QRS morphology that is consistent with the Wolff-Parkinson-White pattern rather than a left bundle branch block. At our initial evaluation, echocardiography demonstrated normal left ventricular function, and his electrocardiogram demonstrated normal sinus rhythm with intermittent complete heart block and ventricular preexcitation suggestive of a mid-septal accessory pathway (Fig. 2). Permanent pacemaker placement was indicated in the setting intermittent atrioventricular nodal and accessory pathway conduction. Given the patient's symptomatic high-grade atrioventricular block, it was prudent to proceed with pacemaker placement prior to electrophysiology study and accessory pathway ablation because of the hypothesised mechanism of atrioventricular conduction injury. The patient successfully underwent placement of a dual chamber pacemaker, which led to the resolution of his symptoms. Subsequently, the patient underwent an electrophysiology study which confirmed the presence of a low-risk, mid-septal accessory pathway with no retrograde conduction and no inducible supraventricular tachycardia. The antegrade accessory pathway effective refractory period was noted to be 380 ms at baseline and 310 ms with high-dose isoproterenol infusion. The pathway was determined to be low risk, and no ablation was performed. At the initial pacemaker interrogation, there was an 11% ventricular pacing burden. By follow-up one month later, the pacing burden was 25% and progressed to 96% one year later. This supports that the accessory pathway was spontaneously deteriorating which led to the patient's presentation 10 years after the iatrogenic injury to his atrioventricular node. At three-year follow-up, the patient reported doing well with no recurrence of symptoms. An electrocardiogram at that time showed intermittent pacing consistent with his device interrogations.

Discussion

Wolff-Parkinson-White pattern is defined by the manifestation of preexcitation on an electrocardiogram caused by an atrioventricular accessory pathway bypassing the atrioventricular node. The prevalence of Wolff-Parkinson-White pattern is estimated at 0.2% of the general population with 65% of adolescents and 40% of young adults remaining asymptomatic.[1–3] Young patients with Wolff-Parkinson-White pattern are at a greater risk of developing life-threatening sentinel symptoms compared with adults.[4] Accessory pathways are associated with sporadic development, a familial form due to a mutation in the PRAKAG2 gene, Ebstein's anomaly, genetic hypertrophic cardiomyopathy, and certain metabolic cardiomyopathies such as Pompe disease and Danon disease.[5] Complete atrioventricular block is associated with congenital heart disease (heterotaxy with accompanied atrioventricular canal defects and L-transposition of the great arteries), myocardial infarction, idiopathic fibrosis

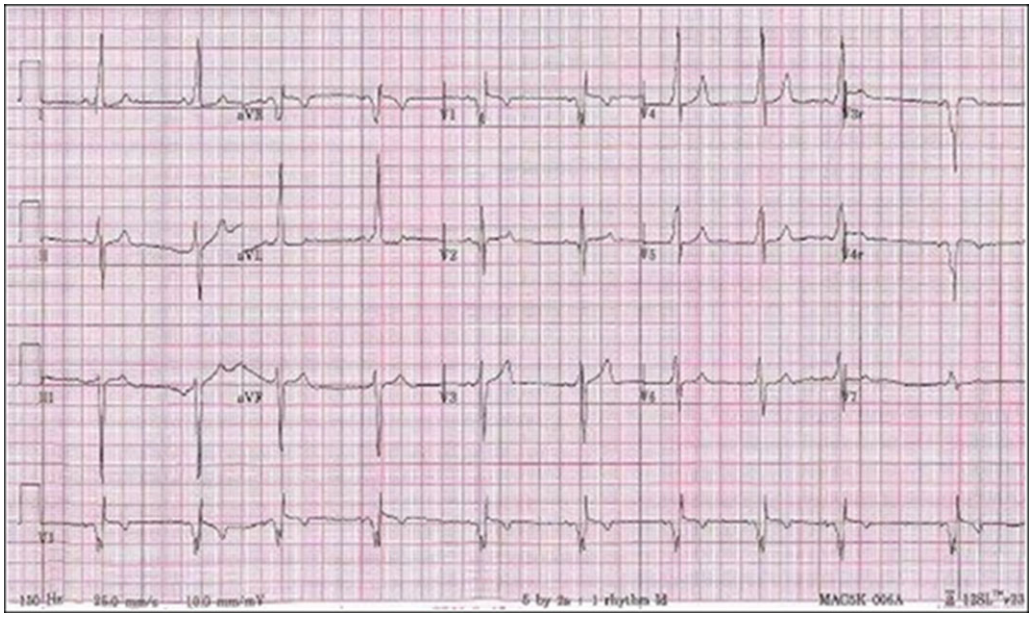

Figure 1. An electrocardiogram which was obtained at a routine follow-up two years prior to our evaluation and was consistent with his postoperative findings.

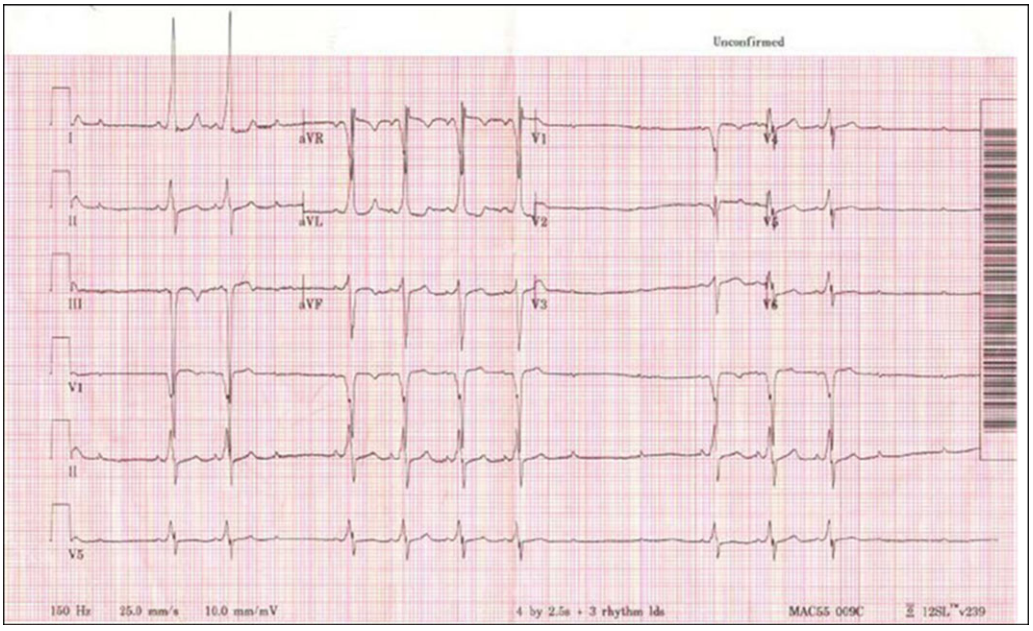

Figure 2. An electrocardiogram obtained during our initial evaluation that demonstrates normal sinus rhythm with complete heart block and ventricular preexcitation suggestive of a mid-septal accessory pathway.

(Lenegre disease) or degradation of the conducting system due to neuromuscular (muscular dystrophy, Kearns-Sayre syndrome) or neoplastic (primary cardiac lymphoma) disorders, infiltrative (sarcoidosis, amyloidosis) and inflammatory myocardial disease, and iatrogenesis due to ablation or surgical complication. Symptoms of an accessory pathway are typically due to tachyarrhythmias but can rarely be secondary to bradyarrythmias as in this case. In the setting of complete atrioventricular node blockade, an accessory pathway has the potential to offer an alternative method of atrioventricular conduction if it is durable, conducts antegrade, and has a suitable refractory period. Symptoms can still develop in one-to-one accessory pathway conduction due to rate-dependent conduction abnormalities or the development of conduction delays. Atrioventricular pathways may spontaneously degrade and disappear.[6] We describe a case of a child with an atrioventricular accessory pathway that bypassed a non-functional atrioventricular node and began to degrade leading to symptomatic intermittent complete heart block.

This case demonstrates the coexistence of presumed iatrogenic complete atrioventricular nodal block with failing antegrade accessory pathway conduction. This is a rare occurrence, and case report observations have described constant or transient

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atrioventricular block in patients with Wolff-Parkinson-White syndrome in the setting of congenital and iatrogenic causes of atrioventricular nodal block.[7] The presence of an accessory pathway can be advantageous in the setting of complete atrioventricular nodal block by providing physiologic one-to-one atrioventricular conduction.[8] Occasionally, an accessory pathway can manifest long after a complete atrioventricular node block is diagnosed and lead to the development of one-to-one atrioventricular conduction without retrograde conduction.[8] Alternatively, accessory pathway conduction can appear at different developmental stages in childhood and adolescence and can exist into adulthood without complication.[9]

An early clue that there had been atrioventricular node damage during the patient's surgery was the postoperative electrocardiogram change. At the time, the patient was described as having a new left bundle branch block, which can sometimes mimic the Wolff-Parkinson-White pattern as seen in Figure 1, which shows a short PR interval, delta wave, and QRS morphology inconsistent with the left bundle branch block. While atrioventricular conduction block can be associated with ostium primum atrial septal defects, our patient had new evidence of conduction delay on postoperative and follow-up electrocardiograms eight years later leading us to suspect an iatrogenic etiology. We hypothesise that the surgery unmasked the Wolff-Parkinson-White pattern. The pathway was most likely concealed preoperatively due to competitive conduction through the atrioventricular node, which led to the pathway being revealed when the atrioventricular node was injured.

Our patient had a non-functional atrioventricular node and an intermittently conducting accessory pathway, the durability of which was unpredictable and required permanent pacemaker placement. Intermittent failure of accessory pathway conduction may manifest as episodic light-headedness, fatigue, exertional intolerance, or syncope in those with atrioventricular nodal disease, and this may be the first clue to the presence of the underlying atrioventricular nodal disease.[10] Others have described presentations of syncope that was initially thought to be due to a tachyarrhythmia associated with an accessory pathway which was revealed to be secondary to complete infrahissian atrioventricular block.[11] Such cases along with ours highlight the importance of perioperative electrocardiogram monitoring and a thorough understanding of a patient's conduction system prior to performing an accessory pathway ablation, which would have left this patient without atrioventricular conduction if performed prior to pacemaker implantation. Regardless of the electrophysiological properties of the atrioventricular accessory pathway, a permanent pacemaker is indicated whenever a high degree atrioventricular node disease is identified because of the propensity of accessory pathways to degrade leading to complete atrioventricular conduction blockade.[12]

This is a rare instance of Wolff-Parkinson-White syndrome presenting with symptoms of bradyarrhythmia rather than tachyarrhythmia. When a previously concealed accessory pathway is unmasked, it is important to consider the reason for the change. This case demonstrates the propensity of accessory pathways to spontaneously degrade and reveal underlying atrioventricular node disease, and for this reason, permanent pacemaker placement is indicated for all high-degree atrioventricular node blocks, regardless of the presence of one-to-one atrioventricular conduction through an accessory pathway. Finally, this case demonstrates the importance of a thorough electrophysiology study prior to accessory pathway ablation.

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Competing interests. None.

Ethical standard. No human or animal experimentation was conducted.

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