A Boy with Fatigue and Heart Block: What's the Mechanism?

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BACKGROUND

Wolff Parkinson White (WPW) pattern is a condition defined by the manifestation of preexcitation on an ECG. This condition is caused by an atrioventricular accessory pathway connecting the atria and ventricles and bypassing the AV node. Such accessory pathways can be clinically silent or be symptomatic leading to the diagnosis of WPW Syndrome. The characteristic of these accessory pathways vary greatly in location and conduction pattern. Some pathways conduct at a resting heart rate while others are only able to be identified while in sinus tachycardia or a supraventricular tachycardia associated with WPW syndrome. Still others are only detectable on an electrophysiology study (EPS). Atrioventricular pathways are often not durable and may spontaneously degrade and disappear.[1]

In the setting of complete AV node block, an accessory pathway has the potential to offer an alternative method of AV conduction. Despite an accessory pathway with 1:1 conduction in the setting of complete heart block, symptoms can still develop due to rate depended conduction abnormalities or the development of conduction blocks in the accessory pathway.

We describe a case of a child with an atrioventricular accessory pathway which bypassed a non-functional AV node and which began to degrade leading to symptoms of intermittent complete heart block.

CASE PRESENTATION

An 11-year-old boy was evaluated for fatigue and decreased exercise tolerance over the past six months prior to presentation. His past medical history was only significant for a surgical repair of an atrial septal defect (ASD) and cleft mitral valve which was performed at 9 months of age at an outside institution using a single pericardial patch. Postoperatively, the patient was described as having a left bundle branch block QRS pattern which was not described previously. Figure (A) shows an an electrocardiogram (ECG) which had been obtained at a routine follow up two years prior to our evaluation and was consistent with his post-operative findings.

Figure (B) shows an ECG obtained during our initial evaluation. It demonstrates normal sinus rhythm with complete heart block and ventricular preexcitation suggestive of a mid-septal accessory pathway.

EVALUATION AND MANAGEMENT

Given the likely need for permanent pacing, it was felt that permanent pacemaker placement was necessary before EP study and ablation. The patient underwent placement of a dual chamber pacemaker for symptomatic high grade atrioventricular block. The patient was then referred for an electrophysiology study which confirmed the presence of a low risk antegrade only mid-septal accessory pathway with no inducible supraventricular tachycardia. Antegrade accessory pathway ERP was noted to be 380 ms at baseline and 310 ms with high dose isopel infusion. At initial pacemaker interrogation, there was an 11% ventricular pacing burden. By follow-up at one month, the pacing burden was 25%, suggesting that the accessory pathway was failing. Clinically, the patient's symptom of fatigue had significantly improved. Figure (C) shows an ECG obtained three years later which reveals intermittent pacing.

DISCUSSION

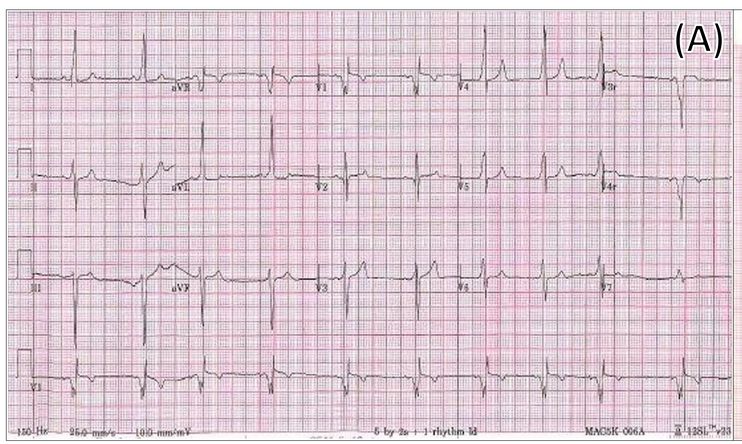
This case demonstrates the coexistence of presumed surgical complete AV nodal block with failing antegrade accessory pathway conduction. This is a rare occurrence and case report observations have described constant or transient AV block in patients with WPW in the setting of congenital and iatrogenic causes of AV nodal block.[2] The presence of an accessory pathway can be advantageous in the setting of complete AV nodal block providing physiologic 1:1 AV conduction.[3] Occasionally, an accessory pathway can develop long after complete AV node block is diagnosed and lead to the development of 1:1 AV conduction without retrograde conduction.[3] Alternatively, accessory pathway conduction can appear at different developmental stages in childhood and adolescence and can exist into adulthood without complication.[4] Alternatively, patients may manifest symptomatically with lightheadedness, fatigue, exertional intolerance, syncope, or seizure when there is interruption in conduction down the accessory pathway, and this may be the first manifestation of AV nodal disease.[5]

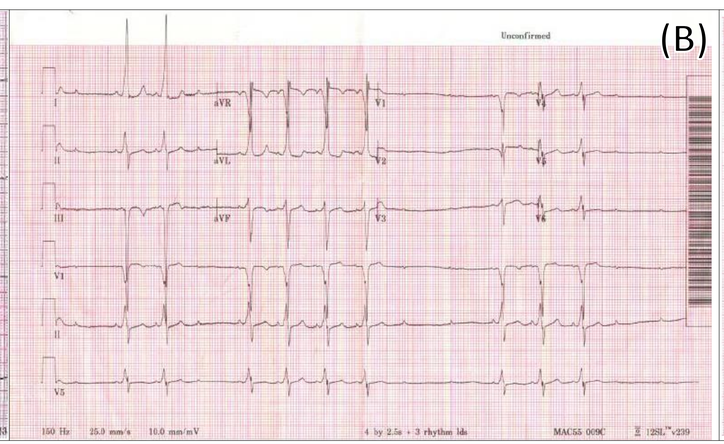
Others have described presentations of syncope that was initially thought to be due to a tachyarrhythmia associated with an accessory pathway which was revealed to actually be secondary to complete infrahissian atrioventricular block. Such cases along with ours demonstrate the importance of a thorough EPS prior to ablation of accessory pathways.[6]

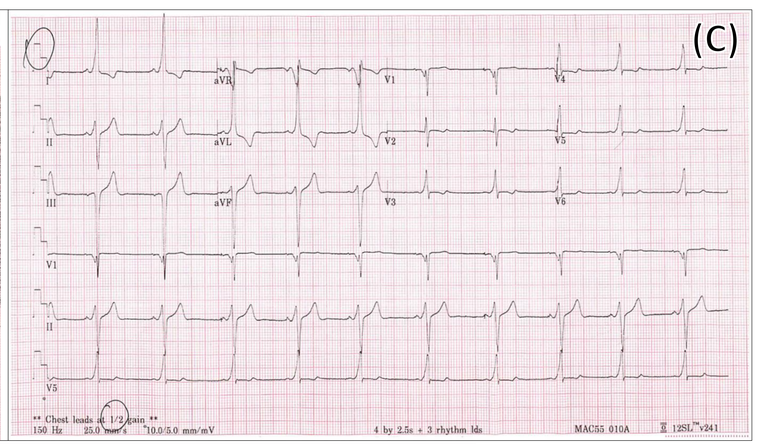
Regardless of the electrophysiological properties of the atrioventricular accessory pathway, a permanent pacemaker is indicated whenever complete heart block is identified because of the propensity of accessory pathways to degrade and developed simultaneous conduction blockade.[7]

Conclusion

In our patient, complete AV nodal block was present most likely as a result of the surgical ASD repair with subsequent dependent accessory pathway conduction. Prior to surgery, there was no documented pre-excitation, suggesting that the antegrade pathway conduction was initially weak. After the AV node was damaged, the antegrade accessory pathway conducted without suppression from more rapid AV nodal conduction. Accessory pathway conduction can have deleterious effects in the presence of atrial fibrillation, but in this case pathway conduction was not sufficiently robust to present a danger. In the absence of AV nodal conduction and with only one accessory pathway without retrograde conduction, there was no potential reentry circuit and this supports why the patient had been asymptomatic until presentation.







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