Epicardial left atrial appendage and biatrial appendage accessory pathways

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BACKGROUND Acute success rates of accessory pathway ablation for Wolff-Parkinson-White (WPW) syndrome can exceed 95%, with rare failures attributed to anatomically complex epicardial connections. Right atrial appendage to right ventricle pathways have been reported, but their left-sided counterparts have only recently been described.

OBJECTIVE The purpose of this study was to report three unique cases of WPW syndrome in children with left atrial appendage and biatrial appendage connections.

RESULTS Three young patients with high-risk accessory pathways (accessory pathway effective refractory period = 190–240 ms) had unsuccessful endocardial ablations despite aggressive efforts with various catheter techniques. One patient had a left atrial appendage to left ventricular connection; the other two had biatrial appendage pathways connected to their respective ventricular surfaces. The latter two patients had a history of ventricular fibrillation: one experiencing ventricular fibrillation in the electrophysiology laboratory and the other suffering from ventricular fibrillation arrest at home. All three patients were taken to the operating room, where the appendages were noted to be diffusely adherent to their ventricles by fibrofatty connections. Dissection of the appendages led to loss of preexcitation and no further tachycardia.

CONCLUSION Surgical management of atrial appendage accessory pathways should be considered if aggressive attempts at endocardial ablation have failed.

KEYWORDS Accessory pathway; Arrhythmia surgery; Atrial appendage; Catheter ablation; Pediatrics; Wolff-Parkinson-White syndrome

ABBREVIATIONS **AP** = accessory pathway; **ERP** = effective refractory period; **LAA** = left atrial appendage; **LV** = left ventricle; **ORT** = orthodromic reciprocating tachycardia; **RAA** = right atrial appendage; **RF** = radiofrequency; **RV** = right ventricle; **VF** = ventricular fibrillation; **WPW** = Wolff-Parkinson-White

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Introduction

Acute success rates for catheter ablation of accessory pathways (APs) can exceed 95% at experienced centers, making it the treatment of choice for most patients with Wolff-Parkinson-White (WPW) syndrome.[1,2] Rare failures are often attributed to anatomically complex APs with epicardial locations. The posterior septal space (where muscular bands or diverticula can span the atrioventricular groove around the proximal coronary venous system) is the most notorious site in this regard.[3] Less commonly, epicardial APs in otherwise normal hearts can involve connections between the right atrial appendage (RAA) and the anterior right ventricle (RV). Milstein et al[4] described such a pathway in 1997, and subsequent reports confirmed similar connections.[5-8] Until recently, there were no reports of APs from the left atrial appendage (LAA) to the left ventricle (LV) other than the speculation of Ohnell[9] in 1943, who described autopsy material from a patient with paroxysmal tachycardia that revealed ". . . a muscle band . . . connecting the left auricle with the left ventricle . . . visible to the naked eye in microscopic preparations." Proof that the LAA can form a functional AP to the LV was finally offered by DiBiase et al,[10] who described two adult patients with APs involving the LAA that were difficult to ablate with conventional catheter techniques. We now report three unusual pediatric patients with high-risk WPW syndrome in whom we verified LAA to LV APs that were not amenable to aggressive efforts with catheter ablation, ultimately requiring surgical intervention. In two of the three patients, concomitant RAA connections were present. These rare cases expand the experience with atypical epicardial APs and provide new surgical observations on the nature of the muscular connections.

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Case reports

Patient 1

A 10-year-old boy with aortic coarctation underwent an uncomplicated surgical repair on day 5 of life by end-to-end

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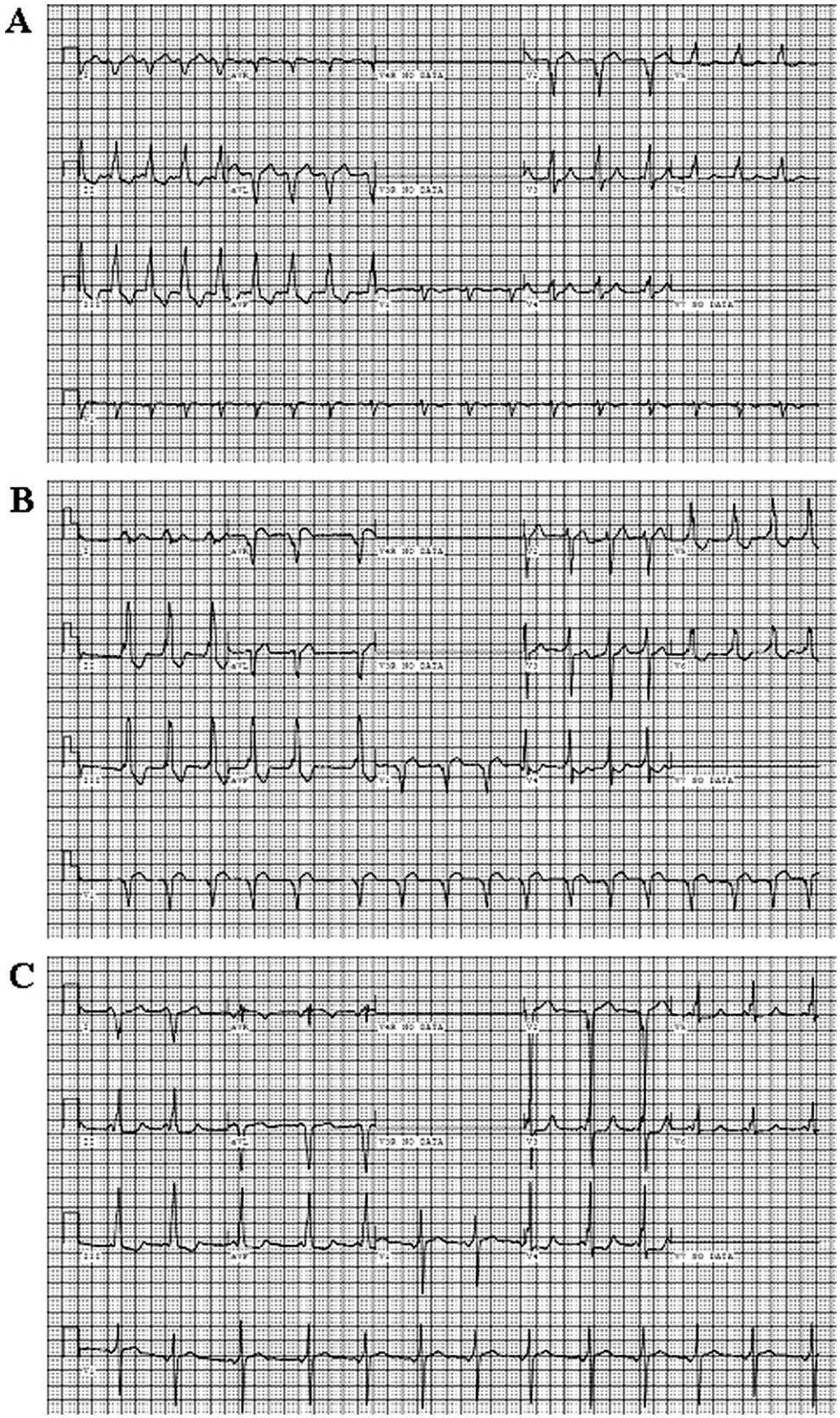

Figure 1 Baseline ECG for patients 1 **(A),** 2 **(B),** and 3 **(C)** demonstrating sinus rhythm with manifest preexcitation.

anastomosis via a left thoracotomy, without entry into the pericardial space. He had manifest preexcitation on ECG (Figure 1) and recurrent episodes of orthodromic reciprocating tachycardia (ORT) beginning prior to surgery. He remained free of tachycardia on beta-blocker throughout infancy. At 20 months (13 kg), he underwent cardiac cath-