A 2-year-old child with coronary sinus diverticulum and Wolff–Parkinson–White syndrome

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Abstract A 2-year-old child having Wolff–Parkinson–White syndrome presented with recurrent drugrefractory tachycardia episodes. On electrophysiological analysis, a coronary sinus diverticulum was discovered. The accessory pathway was successfully eliminated by radiofrequency ablation within the diverticulum.

Keywords: Paediatric arrhythmias; accessory pathway; radiofrequency ablation

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A TRIOVENTRICULAR RE-ENTRY TACHYCARDIA mediated by an accessory pathway is the most common form of tachyarrhythmia in infants and young children. However, drug-refractory Wolff–Parkinson–White syndrome, in particular that associated with coronary sinus diverticulum, is rarely reported in the paediatric age group. Here, we report such a case with successful management.

Case report

A 2-year-old boy weighing 11 kilograms experienced seven episodes of narrow QRS tachycardia since birth. The rate of tachycardia was 250-270 minutes and was associated with listlessness and cold extremities indicating poor perfusion. All episodes needed intravenous administration of adenosine for termination. The electrocardiogram showed sinus rhythm with a negative delta wave in the inferior leads, suggesting a posteriorly located accessory pathway. The clinical examination and echocardiogram were normal. The child was initially started on oral propranolol, but in view of recurrence of tachycardia this was changed to sotalol. Even on a high dose of sotalol (15 milligrams per kilogram per day) he continued to have tachycardia and was referred to our institute for consideration of radiofrequency ablation.

In view of recurrent refractory tachycardia episodes, after discussion with the parents, a permanent cure by radiofrequency ablation was decided upon. An electrophysiological analysis was undertaken under general anaesthesia after stopping sotalol for 48 hours. Bilateral femoral venous access was obtained with two catheters placed from the right and one catheter placed from the left femoral vein. A 4-French deflectable decapolar catheter (Irvine Biomedical, Inc., St. Jude Medical Company, Irvine, CA, USA) was positioned in the coronary sinus and a second 4-French quadripolar catheter was placed in the His bundle position. Maximal preexcitation during the study showed negative delta wave in leads II, III, aVF and V1; there was a deep S wave in V6 and positive delta in aVR (Fig 1). The latter two findings suggested an epicardial accessory pathway. Hence, a coronary sinus angiogram was performed, which showed a diverticulum (Fig 2). Orthodromic atrioventricular re-entrant tachycardia could be easily induced by atrial extra stimuli. Mapping in the neck of the diverticulum showed the earliest ventricular activation in sinus rhythm and the earliest atrial activation during tachycardia. Radiofrequency energy given at this site resulted in elimination of the accessory pathway within 1 second. The radiofrequency energy was given at 15 Watts, temperature attained was 55°C, and the energy was continued for 30 seconds. After this there was no retrograde conduction through the accessory pathway. Following this, vigorous stimulation with isoprenaline did not induce any tachycardia,

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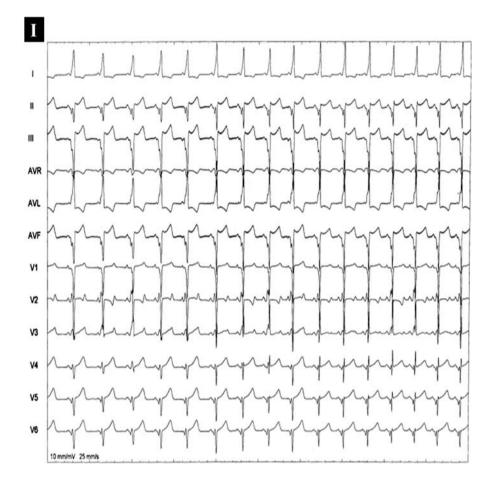


Figure 1.

Electrocardiogram showing maximal pre-excitation.

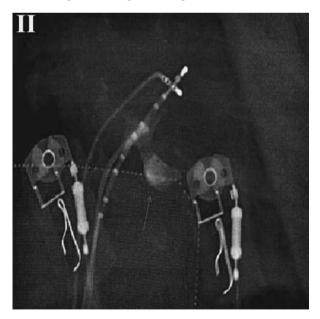


Figure 2. Coronary sinus venogram showing a diverticulum (arrow).

and intravenous administration of adenosine produced transient atrioventricular block. At 4-month follow-up the child was asymptomatic, was not on any medications, and had no pre-excitation in the electrocardiogram.

Discussion

The treatment of choice in older children with atrioventricular re-entry tachycardia is radiofrequency ablation. In infants, however, risk of recurrence is low with only 30–40% of those with onset in infancy having recurrence after infancy.^{1–3} Hence, in infants with atrioventricular re-entry tachycardia, pharmacotherapy is the preferred option. Beyond the age of 1 year, radiofrequency ablation is to be considered when tachycardia episodes occur despite anti-arrhythmic drugs. In our patient this occurred despite supra-therapeutic doses of sotalol, which is a potent anti-arrhythmic agent acting on both antegrade and retrograde accessory pathway conduction.

Coronary sinus diverticula are outpouchings of the coronary sinus, which are rare. In a large series of 480 patients with a posteroseptal or left posterior accessory pathway, Sun et al⁴ demonstrated a coronary sinus diverticulum in 36 patients (7.5%). The diverticula contain myocardial fibres in continuity with the coronary sinus, which connects to both the atria and

ventricles, serving as an accessory pathway. There are only isolated reports of coronary sinus diverticula being diagnosed in children with Wolff–Parkinson–White

only isolated reports of coronary sinus diverticula being diagnosed in children with Wolff–Parkinson–White syndrome.⁵ The diverticulum typically has a narrow neck where the accessory pathway can be targeted for ablation, as in our patient.

The special requirements, especially in very young children, include the need for general anaesthesia and smaller catheters. One of the major concerns of radiofrequency ablation in paediatric patients is the potential for radiofrequency lesions to grow with time.⁶ The other important issue in our case pertains to ablation within the coronary sinus, where the impedance is generally higher (up to 200 Ohms). Thus, to avoid excess heating, coagulum formation and perforation, either temperature control should be used or power should be set initially at very low levels between 3 and 10 Watts.

In summary, this report is a challenging case of successful radiofrequency ablation within the coronary sinus diverticulum in a very young patient.

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