CASE REPORT

Sudden death in a child with an unusual accessory connection

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An 8-year-old boy with Wolff-Parkinson-White syndrome died suddenly whilst exercising. He had been well with no immediate preceding symptoms of an arrhythmia, and was receiving no drugs. As an infant he had suffered recurrent episodes of atrioventricular reciprocating tachycardia (AVRT) and had documented pre-excited atrial fibrillation. At electrophysiological (EP) study, the refractory periods of his two accessory pathways were less than 220 ms. Drugs were discontinued at 4 years of age when he remained asymptomatic. Autopsy confirmed a very unusual accessory pathway. It was a muscular connection between the base of the right atrial appendage and the right ventricle, distant from the annulus of the tricuspid valve.

Key Words: Wolff-Parkinson-White syndrome, sudden death, childhood, accessory connection.

Introduction

Sudden death in Wolff-Parkinson-White syndrome is primarily thought to occur when atrial fibrillation conducts rapidly to the ventricle in the setting of a pathway with a short refractory period. The true incidence of sudden death in Wolff-Parkinson-White syndrome in childhood is unknown. The majority of studies have highly selected patients from tertiary referral centres. In population studies of Wolff-Parkinson-White syndrome, sudden death is rare, particularly in the paediatric age group. We report the case of an 8-year-old boy who died suddenly with Wolff-Parkinson-White syndrome. At autopsy an unusual accessory connection from the base of the right atrial appendage to the right ventricle was documented.

Case history

A male infant of 10 days of age was transferred to our hospital. At 1 week of age he was admitted to his local hospital, tachypnoeic, irritable, reluctant to feed and was found to have a regular wide complex tachycardia (see Fig. 1). Vagal manoeuvres, facial ice and intravenous adenosine had no effect and intravenous flecainide slowed the tachycardia without termination. Sinus rhythm was restored with DC cardioversion. The electrocardiogram was consistent with Wolff-Parkinson-White syndrome. The vector of the delta wave suggested a right-sided pathway.

At 6 weeks the infant was readmitted with repeated episodes of wide complex tachycardia. Flecainide was discontinued for diagnostic EP study. Prior to the study, the infant developed an irregular wide complex tachycardia with a minimum R-R interval of 220 ms. At EP study, right atrial, right ventricular, and His bundle electrograms were recorded. A mapping catheter was placed in the left ventricle using the retrograde aortic approach. AVRT and atrial tachycardia (with 1:1 conduction to the ventricle) was easily inducible with programmed atrial stimulation. Two accessory connections were identified: right and left free wall. Both had an anterograde refractory period of <220 ms.

The infant was started on oral amiodarone with good clinical effect. He sustained one further episode of AVRT at 10 months of age. Digoxin was added to the amiodarone and the child remained well. At 3 years of age he remained well.
age his digoxin was discontinued, as was the amiodarone at 4 years of age. He remained symptom free although electrocardiograms demonstrated persistent pre-excitation. At the age of 8 years he died suddenly whilst riding his bicycle with no prior warning symptoms. Attempts at resuscitation failed.

At autopsy no cause for death other than arrhythmia could be found. At gross examination the cardiac chambers were found to be arranged in the usual fashion, with normal connections and coronary arteries. On dissecting into the right atrioventricular groove, the right atrial appendage was resistant to being lifted away from the ventricular mass. Close inspection revealed a band-like muscular structure extending from the underside of the atrial appendage to the right ventricle [see Fig. 2(a)]. Internally this structure corresponded to a pouch with an orifice of 5 mm in the right atrial appendage, hidden between the pectinate muscles [see Fig. 2(b)]. The atrioventricular conduction system was examined in detail histologically, including sections of the right AV junction. The atrioventricular node and proximal portions of the atrioventricular conduction system were normal in location and configuration. There were multiple venous channels on the underside of the right atrial appendage. A pouch was seen in the floor of the atrial appendage 8 mm from the septum. It was ultimately traced to the right ventricular wall in continuity with one of the small veins. The pouch had a muscular wall that continued into the ventricular myocardium [see Fig. 2(c, d)] approximately 5 mm away from the annular insertion of the tricuspid valve. The atrial vestibular wall at this level was separated from the area of atrioventricular muscular continuity by an extensive atrioventricular groove [see Fig. 2(c)]. Thus a right-sided pathway was identified straddling between the base of the right atrial appendage and ventricle, through the epicardial fat, remote from the annulus of the tricuspid valve. The left free wall accessory pathway was not identified by gross examination. Detailed serial sectioning of the left AV groove was not undertaken.

Discussion

Early series of symptomatic Wolff-Parkinson-White syndrome in infancy emphasized the spontaneous resolution of arrhythmia, although the majority of children remained on antiarrhythmic medication[3,4]. In a series of 140 children with Wolff-Parkinson-White syndrome and an initial episode of AVRT before the age of 18 years, over half presented in the first year of life. In 93% of these cases, tachycardia disappeared after 1 year, but reappeared in 31% at an average of 8 years. This appeared to be independent of drug therapy. If tachycardia occurred after the age of 5 years, it tended to persist[5]. This natural history has led to the commonly adopted policy of withdrawing drug therapy after infancy and reinstituting therapy with symptomatic recurrence[6].

Figure 1  12 lead electrocardiogram (25 mm . s⁻¹) of the presenting tachycardia at 1 week of age.
Sudden death is the most dramatic and worrying complication of Wolff-Parkinson-White syndrome; however, it is uncommon in community studies\[2\]. Information on the clinical and electrophysiological parameters assessing the risk of ventricular fibrillation have largely been gathered from case series of adult patients resuscitated from sudden cardiac death referred to tertiary care centres. The most sensitive markers are the anterograde conduction properties of the pathway, particularly the shortest pre-excited R-R interval in atrial fibrillation. Clinical markers include multiple accessory pathways, septal pathways, and multiple types of clinical tachycardia. However, these findings are not specific and have a low predictive value\[1,7,8\]. A series of 60 children with symptomatic Wolff-Parkinson-White syndrome who had comprehensive electrophysiological evaluation were divided into high risk (10 with a history of cardiac arrest), intermediate risk (19 with a history of atrial fibrillation or syncope), and low risk (31 with orthodromic AVRT). There was no difference between the groups in terms of age, duration of symptoms, incidence of congenital heart disease, the presence of multiple pathways or accessory pathway site. The most sensitive marker of risk was a shortest pre-excited R-R interval of <220 ms, but this also occurred in 35% of the low-risk group\[9\].

This child presented in 1990, prior to the widespread use of radiofrequency ablation. At that time our institution was not undertaking ablation in infancy or early childhood, but we would now consider early ablation in such a patient. When the patient became asymptomatic after infancy, a conservative approach was adopted with a view to definitive therapy with recurrence of symptoms (in the belief that sudden death in early childhood is rare). However as ‘high-risk’ markers were identified in early life, we feel it would have been prudent to continue medical therapy (unless further EP study demonstrated modification of the anterograde conduction properties of the accessory pathways).

Free wall accessory pathways are typically located close to the hinge of the valve in location close to the endocardial aspect. The majority of reported epicardial accessory pathways are adjacent to a coronary sinus diverticulum, the middle cardiac vein, or the great cardiac vein\[10\]. Pre-excitation and neonatal supraventricular tachycardia has been reported in an unusual case of a giant right atrial diverticulum that extended from the lateral right atrial wall to the right ventricular apex\[11\]. Milstein et al\[12\] have also reported an unusually located accessory connection localized to the right atrial appendage. Following multiple attempts at catheter ablation, surgery was successful when a bridge of tissue (that traversed epicardially from the base of the right atrial appendage into the fat pad overlying the base of the right ventricle) was transected. This resulted in abrupt loss of anterograde and retrograde conduction. Soejima et al\[13\] also report an accessory pathway successfully ablated in the right atrial appendage. Clues that this unusual pathway may be present include earliest atrial activation at the appendage during orthodromic AVRT, absence of an accessory pathway potential, and lack of transient interruption during ablation at the site of earliest activation at the annulus. In this case the RA appendage should be mapped. There are few
opportunities for pathological study of accessory pathways in children. Our patient is the first where this unusual pathway has been histologically documented. The anatomical understanding of such pathways is important when undertaking ablation.

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References

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