

SCIENTIFIC LETTER

Asymptomatic ventricular pre-excitation in children and adolescents: a 15 year follow up study

B Sarubbi, G Scognamiglio, G Limongelli, B Mercurio, G Pacileo, C Pisacane, M G Russo, R Calabrò

Heart 2003;89:215–217

The term Wolff-Parkinson-White (WPW) syndrome refers to people with a ventricular pre-excitation (VPE) pattern on ECG who experience symptomatic bouts of tachycardia.¹ Asymptomatic patients with VPE are better described as simply having a “VPE pattern”.

Diagnostic assessment and treatment have been described in detail in patients with symptomatic WPW syndrome, but little information exists about the prognosis of VPE discovered on routine ECG in an asymptomatic person.

Our study aimed to examine retrospectively the follow up of a cohort of children with asymptomatic VPE, referred during a 15 year period to our unit.

METHODS

From January 1985 to July 2001, 98 children (56 male/42 female) found incidentally to have a VPE on routine ECG were referred to our division. All patients underwent a complete evaluation. The patients were considered asymptomatic if they had no documented tachycardia and no history of palpitations. At recruitment, all the patients underwent clinical examination, ECG, echocardiogram, ECG Holter, and, when possible, exercise testing. Accessory pathway location was established by the use of ECG criteria according to Fitzpatrick's algorithm.² The start of the follow up period dated from the time when the first ECG with a δ wave was recorded.

Intermittence of VPE was defined as reappearance after loss. Loss of VPE was defined as the absence of VPE for more than three consecutive controls.

In the absence of symptoms, patients were seen in outpatient clinic at 6–12 month intervals.

Electrophysiological study (EPS)

The shortest cycle length showing pre-excitation was designated as the minimal cycle length that maintained 1:1 conduction over the accessory pathway. Programmed atrial stimulation at a basic cycle length of 600, 500, and 400 ms with the introduction of one and two extrastimuli was performed. The effective refractory period of the accessory pathway was the longest A1–A2 interval without VPE. If atrial fibrillation (AF) was not initiated with programmed stimulation, attempts were made to induce AF by repeated 15 second bursts at a cycle length of 300–100 ms. Isoproterenol (0.02–1 g/min) was then infused to increase the sinus rate at least 130 beats/min and the pacing protocol was repeated. Non-sustained AF or reciprocating tachycardia was defined as that arrhythmia that terminated spontaneously within 30 seconds. Atrial vulnerability was defined as the induction of sustained AF (≥ 30 seconds) by programmed stimulation or incremental atrial pacing. Standard measurements recorded during AF included the minimal RR interval between two consecutive pre-excited complexes and the average RR interval of all cycles.

RESULTS

Among the patients found to have a VPE, 57 (35 male/22 female) entered the study, as they were completely asymptomatic at diagnosis. The mean (SD) age at recruitment was 9.7 (5.4) years (range 1 month to 17 years). Of the 57 patients recruited, 36 (63.1%) were first diagnosed at study entry.

Congenital heart disease

Eleven patients (19.2%) exhibited congenital heart disease together with VPE (3 atrial septal defects, 2 mitral valve prolapse, 1 aneurysm of the oval fossa, 1 atrioventricular canal, 1 patent ductus arteriosus, 1 subaortic stenosis, 1 congenitally corrected transposition of the great arteries, 1 double outlet right ventricle).

Clinical course

Based on a mean follow up period of 48 months, the cumulative observation period was 5.5 patient-years. During the follow up, 7 patients (12.2%), aged 7.7 (2.4) years, were found to have intermittent VPE on ECG, ECG Holter or exercise test, and 3 patients (5.2%), aged 15.3 (8.3) years, were found to have complete loss of VPE. During the follow up 5 patients (8.7%), aged 9.6 (2.9) years, were referred with symptomatic supraventricular tachycardia (SVT) (4 reciprocating tachycardia, 1 AF) and underwent medical treatment or radiofrequency catheter ablation (RFCA).

No patients had SVT induction during exercise test. During the follow up one patient aged 8 years, whose parents refused to perform EPS, died from sudden death. His pathway localisation was right posteroseptal.

Electrophysiological study

For risk stratification an EPS was proposed to all the patients. Thirty five patients, aged 8.9 (4.6) years, underwent EPS, and 17(48.7%), aged 10.2 (4.7) years, experienced sustained SVT. The tachycardia was initiated in the basal state in 13 patients (aged 10.2 (4.5) years) and after isoproterenol in the other 4 patients (aged 10 (3.2) years).

Orthodromic SVT (cycle length 299.2 (51.6) ms, range 210–370 ms) was recorded in 14 patients (8 male/6 female) aged 9.2 (4.6) years (fig 1). In 2 patients, aged 5.5 (0.7) years, both orthodromic and antidromic SVT were recorded, with different cycle lengths (280 (99) ms and 255 (77.8) ms respectively), as expression of multiple pathways. Antidromic SVT (cycle length 240 ms) was recorded alone in only 1 patient aged 8 years. AF was recorded in 4 patients (aged 10.2 (4.3) years). In 2 patients (aged 7.5 (4.9) years) it was

Abbreviations: AF, atrial fibrillation; EPS, electrophysiological study; RFCA, radiofrequency catheter ablation; SVT, supraventricular tachycardia; VF, ventricular fibrillation; VPE; ventricular pre-excitation; WPW, Wolff-Parkinson-White

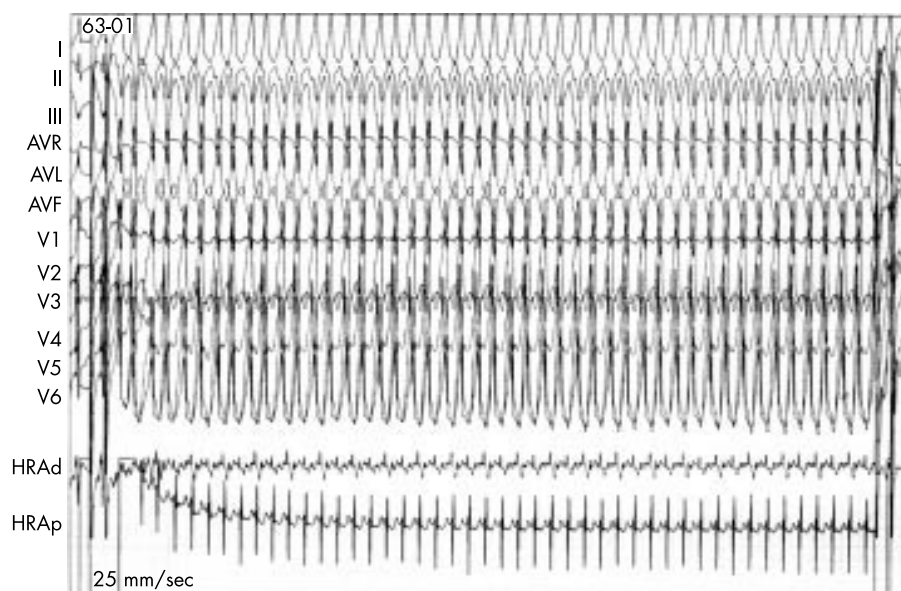


Figure 1 Antidromic supraventricular tachycardia in a patient aged 6 years (cycle length 210 ms).

recorded after the induction of orthodromic or antidromic tachycardia (fig 2). In the other 2 patients (aged 13 (1.4) years) AF was the first and only arrhythmic event. The average RR interval during AF ranged between 230–290 ms. The minimal RR interval between two consecutive pre-excited QRS ranged between 250–230 ms (mean 237.5 (9.6) ms). In the 18 patients (aged 10.1 (5.1) years) that presented no induced SVT in the EPS, the 1:1 conduction over the accessory pathway ranged between 210–350 ms (mean 276.3 (39.1) ms). In the 17 patients (aged 10.2 (4.7) years) with SVT and/or AF in the EPS, the 1:1 conduction over the accessory pathway ranged between 200–320ms (mean 255.3 (27.2) ms). For the patients who sustained SVT, either re-entry tachycardia or AF, during EPS, medical treatment or RFCA were proposed. The patients who did not experience SVT, but had a 1:1 conduction over the pathway \leq 250 ms in the control state and/or \leq 220 ms after isoproterenol (5 patients), were discharged without anti-arrhythmic medication, although they were prohibited from participating in sport.

Accessory pathway location

In 32 patients (aged 10.2 (5.3) years) the pathway localisation, according to Fitzpatrick criteria, was right (2 posterolateral, 10 anterolateral, 9 anteroseptal, 3 midseptal, 8 posteroseptal), and in 25 patients (aged 9 (5.4) years) the pathway localisation was left (14 anterolateral, 8 posteroseptal, 3 posterolateral).

In the 5 patients who presented with clinical tachycardia the pathway localisations were: right anterolateral (2), right anteroseptal (1), left anterolateral (1), and left posteroseptal (1).

In the 14 patients who presented with induced orthodromic tachycardia the pathway localisations were: left (6 anterolateral; 4 male/2 female), and right (3 posteroseptal, 3 anteroseptal, 2 anterolateral; 4 male/4 female).

In the 3 patients who presented with antidromic tachycardia in the EPS, including the 2 patients who presented both antidromic and orthodromic tachycardia, the pathway localisations were: 1 midseptal, 2 left anterolateral.

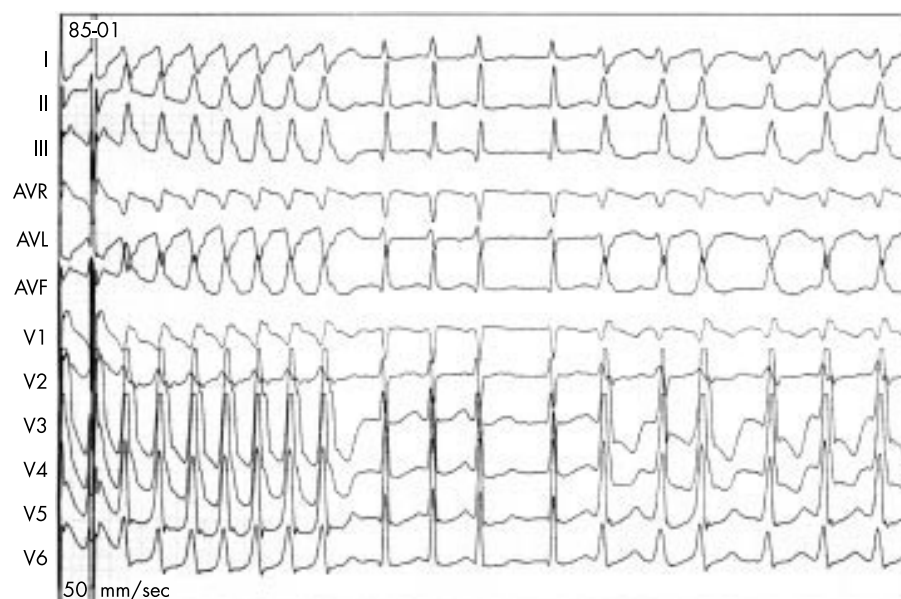


Figure 2 Spontaneous degeneration of previously induced antidromic supraventricular tachycardia.

In the 4 patients who presented with AF during the EPS, the pathway localisations were left in 2 patients (1 anterolateral, 1 posterolateral), and right in 2 patients (1 anterolateral, 1 anteroseptal).

DISCUSSION

Asymptomatic people with VPE are supposed to have a benign prognosis, although it has been shown³ that almost 30% of initially asymptomatic individuals develop symptoms over a 12 year follow up period, and some patients can present with ventricular fibrillation (VF) as the first manifestation of the syndrome.⁴ The presence in our series of one patient, aged 8 years and previously asymptomatic, who died from sudden death, raises the necessity for a better risk stratification of this population.

Electrophysiological study and management

Assessment of sudden death risk is most controversial in the completely asymptomatic individual with WPW pattern. No definite clinical or electrophysiological variables have been found to predict which people are likely to become symptomatic. However, an extremely rapid ventricular response over the accessory pathway during AF, either spontaneous or induced at EPS, has been considered the most striking and consistent electrophysiological finding in the WPW patient resuscitated from VF. The average ventricular rate during induced AF has been found slightly slower than during the spontaneous arrhythmia, but a good linear correlation between the two measurements has validated the utility of observations during the EPS.⁵

In our series almost 49% of the patients who underwent an EPS experienced SVT. The presence of a high proportion of patients in which the tachycardia initiated after isoproterenol infusion (4/17 patients, 23.5%) confirm the necessity for a very aggressive protocol of induction, and perhaps the necessity of contrasting the effect of sedation with the use of adrenergic agents. No differences in sex, age, and pathway localisation were found between patients who had orthodromic, antidromic tachycardia, AF, or no arrhythmia at EPS. Age at study was not related to AF induction, as generally thought. The use of antiarrhythmic drugs or RFCA for patients, previously asymptomatic, who experienced sustained SVT during EPS has been widely debated. No study has been designed to confirm the effective value of this "aggressive" management in the prevention of SD. The decision to prescribe antiarrhythmic drugs or RFCA to all the patients who experienced sustained SVT, and to prohibit patients with a faster 1:1 conduction over the accessory pathway from participating in sport, even in the absence of sustained tachycardia, could be criticised in many ways, but it was considered the only way to prevent those "rare" cases of sudden death.

Pathway location

No particular pathway was found to be associated with spontaneous or induced tachycardia. In our series there was a higher than expected prevalence of right located pathways. However, in children, right sided pathways have been reported to be slightly more common than in adults. Furthermore, the higher prevalence of right located pathways can be explained by the low sensitivity of the surface ECG to localise left free wall pathway, and the possible difficulties in correctly identifying multiple pathways, other than the possible changes over time of the conductivity.⁶

Congenital heart disease

Our series found a higher than expected prevalence of congenital heart disease (almost 20%). However, this higher prevalence may be an inevitable result of bias selection, as these patients first came to our attention because of associated structural heart disease, and VPE was incidentally found as a result of complete diagnostic assessment.

ACKNOWLEDGEMENT

This paper supported by the Institute of Cardiac Researches, Monaldi Hospital, Naples and financed by the Operative Programme Plain "Critical Neonatal Congenital Heart Disease (CCCN5)" BOO6 of University and Scientific Research Ministry (MURST), and the European Community (711/1998). We gratefully thank all the nurses of the Division of Paediatric Cardiology, Monaldi Hospital, for the technical support and cooperation in the outpatient clinic and cath lab, particularly Mr Riccardo Braun, Mrs Agnese Caccese, Mrs Maria Izzo, Mrs Maria Gabriella Lettieri, and Mrs Ada Orefice.

Authors' affiliations

B Sarubbi, G Scognamiglio, G Limongelli, B Mercurio, G Pacileo, C Pisacane, M G Russo, R Calabrò, Second University of Naples, Chair of Cardiology, Division of Pediatric Cardiology, Monaldi Hospital Naples, Italy

Correspondence to: Dr Berardo Sarubbi, Via Tino di Camaino, 23 80128 Napoli, Italy; bsarubb@tin.it

Accepted 25 July 2002

REFERENCES

- 1 Wolff L, Parkinson J, White P. Bundle branch block with short PR interval in healthy young people prone to paroxysmal tachycardia. *Am Heart J* 1930;**5**:685-704.
- 2 Fitzpatrick AP, Gonzales RG, Lesh MD, et al. New algorithm for the localisation of accessory atrioventricular connections using a baseline electrocardiogram. *J Am Coll Cardiol* 1994;**23**:107-17.
- 3 Munger TM, Packer DL, Hammill SC, et al. A population study of the natural history of Wolff-Parkinson-White syndrome in Olmsted County, Minnesota, 1953-1989. *Circulation* 1993;**87**:866-73.
- 4 Klein GJ, Bashore TM, Sellers TD, et al. Ventricular fibrillation in the Wolff-Parkinson-White syndrome. *N Engl J Med* 1979;**301**:1080-5.
- 5 Rinne C, Klein GJ, Sharma AD, et al. Relation between clinical presentation and induced arrhythmias in the Wolff-Parkinson-White syndrome. *Am J Cardiol* 1987;**60**:576-9.
- 6 Sarubbi B, Elizari A, Somerville J. A pre-excitation growing-up over the time. *Heart* 2000;**83**:146.