A novel mechanism of sudden infant death syndrome during atrioventricular reentrant tachycardia: a case report

Hitoshi Mori 1,2, Naokata Sumitomo 1*, Kenta Tsutsui 2, and Taisuke Nabeshima 1

1Department of Pediatric Cardiology, Saitama Medical University International Medical Center, 1397-1 Yamane Hidaka, Saitama 350-1298, Japan; and 2Department of Cardiology, Saitama Medical University International Medical Center, 1397-1 Yamane Hidaka, Saitama 350-1298, Japan

Received 14 November 2021; first decision 9 December 2021; accepted 11 June 2022; online publish-ahead-of-print 16 June 2022

Background Although orthodromic atrioventricular reentrant tachycardia (AVRT) using retrograde conduction through an accessory pathway (AP) is a common manifestation of Wolff-Parkinson-White (WPW) syndrome, a rare yet critical consequence is sudden cardiac arrest in a few patients. This fatal event used to be reported as a result of rapid atrioventricular conduction of atrial fibrillation via an AP.

Case summary A 3-month-old infant with WPW syndrome had AVRT accompanied by global cardiac ischaemia, apparently caused by a rapid ventricular rate itself that degenerated into ventricular fibrillation during the AVRT.

Discussion Our case suggested that orthodromic AVRT may be sufficient to cause WPW-related sudden cardiac death (SCD) or sudden infant death syndrome via fatal ischaemia and ventricular arrhythmias even when the effective refractory period of an antegrade AP conduction is long or even when an antegrade AP is not present. It is possible that an AP ablation in those who have a history of a fast orthodromic AVRT would be useful to prevent SCD in addition to symptom control.

Keywords Case report • Wolff-Parkinson-White syndrome • Sudden cardiac death • Ventricular fibrillation • Supraventricular tachycardia • Sudden infant death syndrome

ESC Curriculum 5.1 Palpitations • 5.5 Supraventricular tachycardia

Learning points
- Global cardiac ischaemia during orthodromic atrioventricular reentrant tachycardia (AVRT) may become a novel mechanism that may lead to sudden cardiac death in patients with WPW syndrome.
- An accessory pathway ablation in those who have a history of fast orthodromic AVRT would be useful to prevent fatal events.
Introduction

Although orthodromic atrioventricular reentrant tachycardia (AVRT) using retrograde conduction through an accessory pathway (AP) is a common manifestation of Wolff-Parkinson-White (WPW) syndrome, a rare yet critically sudden cardiac death (SCD) occurs in 1.5–2.2% of patients.\textsuperscript{1,2} This fatal event used to be reported as a result of rapid atrioventricular conduction of atrial fibrillation (AF) via an AP, so-called ‘pre-excited AF’ or ‘pseudo ventricular tachycardia’. In this context, a short AP antegrade refractory period and the initiation of AF have been reported to be a risk factor of SCD.\textsuperscript{1,3} In other words, a rapid ventricular contraction is the key to the WPW-related fatal events. Although this assumption may be true, the underlying mechanism has been elusive. Here, we report an infant case of WPW syndrome that suggested an AVRT accompanied by global cardiac ischaemia, apparently caused by the rapid ventricular rate itself that degenerated into ventricular fibrillation (VF) during the AVRT.

Timeline

| Before his delivery | His mother had gestational diabetes. He was delivered by a cesarean section at 38 weeks and 3 days. |
|---------------------|-------------------------------------------------------------------------------------------------|
| 15 min after his delivery | Supraventricular tachycardia (SVT) first became documented, and he was diagnosed with WPW syndrome. A rapid infusion of adenosine triphosphate terminated the SVT. |
| 1-day-old to 3-month-old | Propranolol (4 mg/kg/day), sotalol (6 mg/kg/day), and flecainide (200 mg/m\textsuperscript{2}/day) were initiated, but the tachycardia was difficult to control. |
| 3-month-old and 4-days-old | He was admitted to our hospital for radiofrequency catheter ablation. |

Case report

A 3-month-old Asian infant was admitted to our hospital for radiofrequency catheter ablation (RFCA) targeting a drug-resistant AVRT. Although his mother had gestational diabetes, his family had no past medical history of cardiac disease. She was gravida 1 para 1. He was delivered by a cesarean section at 38 weeks and 3 days. He did not have any congenital abnormalities and all the newborn screening blood tests were within normal range. Supraventricular tachycardia (SVT) was first documented 15 min after his delivery. A rapid infusion of adenosine triphosphate (ATP, 0.2 mg/kg) terminated the SVT, and the tachycardia turned out to be AVRT due to ventricular pre-excitation after termination of the SVT. Echocardiography revealed a normal left ventricular function without any structural heart disease. Although the oral administration of propranolol (4 mg/kg/d), sotalol (6 mg/kg/d), and flecainide (200 mg/m\textsuperscript{2}/d) failed to control this tachycardia, the tachycardia occurred one to two times a week. He was referred to our hospital when he was 3 months old. On admission, his body weight and body height were 8.5 kg and 66 cm, respectively. He did not have any heart failure symptoms.

After written informed consent was obtained from the guardians, ablation was performed under general anaesthesia. The baseline electrocardiogram (ECG, Figure 1A) suggested that an AP was located on the left side. Just after a 7Fr long sheath and 5Fr sheath were secured from the right femoral vein and right jugular vein, AVRT (tachycardia cycle length: 232 ms) spontaneously initiated without a catecholaminergic drug infusion (Figure 1B). While preparing the ATP to terminate the SVT, an ECG exhibited progressive horizontal ST-T depression (Figure 1C) with a down sloping of the ST-T depression and reciprocal ST-T elevation in the aVR lead (Figure 1D), which further worsened suggesting global cardiac ischaemia (Figure 1E). Such a sequence culminated in polymorphic premature ventricular contractions (PVCs) and degenerated into VF (Figure 2). Two external cardiac defibrillations (20 J) were required to terminate this VF. The haemodynamic status was stable after defibrillation, and the procedure was restarted.

A 5Fr decapolar catheter (SNAKE®, Japan Lifeline) was positioned in the coronary sinus, and an ablation catheter (THERMOCOOL SF®, Biosense Webster, Irvine, CA, USA) was positioned into the left atrium (LA) via a patent foramen ovale. An electrophysiological study revealed that the earliest antegrade and retrograde conduction were located on the lateral wall of the LA, and the conduction was not interrupted under a bolus infusion of ATP. Atrial programmed stimulation demonstrated that the effective refractory period (ERP) of the antegrade conduction of the AP was 240 ms and did not reveal any decremental property. The earliest ventricular activation site was confirmed on the lateral wall of the LA during sinus rhythm for which radiofrequency current (30 watt, 30 s) was delivered. The AP conduction disappeared immediately after the first burn, and the AP conduction never recurred during a 60-minute waiting period and the procedure was concluded. During 8 months of follow-up, no recurrence of the AVRT was documented.

Discussion

WPW syndrome has a risk of SCD, which is generally considered to be due to VF from rapid atrioventricular conduction of AF through the AP, with a 10-year risk of SCD ranging from 0.15% to 0.24%.\textsuperscript{1,4} Most patients who experience VF are asymptomatic prior to the event, and the shortest pre-excited RR interval during AF is <220 ms, which is believed to be a risk factor of a fatal event.\textsuperscript{1,5} AF triggered by AVRT is reported to be another strong risk factor of VF.\textsuperscript{1} Thus, one may postulate that the AP antegrade conduction causes VF when AF and excessive atrioventricular AP conduction occur simultaneously while the AP retrograde conduction does not. However, the present case clearly demonstrated that orthodromic AVRT, not pre-excited AF with excessive atrioventricular conduction, deteriorated into VF within 12 min. As described above, the progressive ST-T depression and concomitant reciprocal change in the aVR lead suggested broad cardiac ischaemia. Previous reports noted that the retrograde AP ERP and tachycardia cycle length were significantly shorter with a younger age.\textsuperscript{6} This suggested that younger WPW syndrome patients could have a greater risk of global cardiac ischaemia due to a fast tachycardia event than adults.
Figure 1 ST-T change on the baseline electrocardiogram and during the tachycardia. (A) Shows the baseline electrocardiogram. The accessory pathway was suspected to be located in the left atrium. (B) Shows the 12-lead electrocardiogram with a tachycardia cycle length of $232$ ms. (C) Shows the ischaemic changes with horizontal ST-T depression and that the horizontal ST-T depression changed to a down sloping ST-T depression with reciprocal ST-T elevation in lead aVR (Figure 1D and 1E).

Figure 2 Induced ventricular fibrillation before the catheter ablation. Polymorphic premature ventricular contractions occurred and ventricular fibrillation was suddenly induced.
The present study suggested that orthodromic AVRT may be sufficient to cause WPW-related SCD or sudden infant death syndrome via fatal ischaemia and ventricular arrhythmias even when the ERP of the antegrade AP conduction is long, or even when an antegrade AP is not present.

Catheter ablation of the AP is recommended as the first-line therapy in patients who have had AF and/or AVRT as a class I recommendation.7 Drug therapy, drugs such as flecainide or propafenone, can be considered as a class IIa recommendation. The therapeutic concept of orthodromic AVRT focuses on symptom control but does not necessarily focus on primary prevention of SCD via fatal arrhythmias. The concept was based on the research in an adult population. There have been few reports that have focused on the pediatric population. However, our case led us to consider that orthodromic AVRT itself may be sufficient to induce fatal arrhythmias in a small infant. This idea is consistent with the finding of a previous study that a part of not only the symptomatic group but also the asymptomatic group experienced VF.8 It is tempting to speculate that, while symptomatic patients are more likely to be treated, asymptomatic patients are likely to be left untreated with a risk of SCD.

In conclusion, although the incidence of orthodromic AVRT underlying SCD via cardiac ischaemia remains elusive, our case suggested a possibility that an AP ablation for those who have a history of a fast orthodromic AVRT would be useful to prevent SCD in addition to symptom control.

Lead author biography

Dr. Sumitomo graduated from Nihon University School of Medicine in 1981. He was studied abroad in Texas Children’s Hospital, TX from 1989 to 1992. And, he has been working in the Saitama Medical University International Medical Center as a professor of paediatric cardiology since 2014. His expert field is arrhythmia in paediatric, congenital heart disease, and inherited arrhythmia.

Consent: The authors confirm that written consent for submission and publication of this case report including the images and associated text has been obtained from the guardians in line with COPE guidance.

Conflict of interest: None declared.

Funding: None declared.

References

1. Pappone C, Vicedomini G, Manguso F, Saviano M, Baldi M, Pappone A, Ciaccio C, Giannelli L, Ionescu B, Petretta A, Vitale R, Cuko A, Calovic Z, Fundalitis A, Moscatiello M, Tavazzi L, Santinelli V. Wolff-Parkinson-White syndrome in the era of catheter ablation: insights from a registry study of 2169 patients. Circulation 2014;130:811–819.
2. Timmermans C, Smeets JL, Rodriguez LM, Vrouchos G, van den Dool A, Wellens HJ. Aborted sudden death in the Wolff-Parkinson-White syndrome. Am J Cardiol 1995;76:492–494.
3. Etheridge SP, Escudero CA, Blaufax AD, Law IH, Dechert-Crooks BE, Stephenson EA, Dubin AM, Ceresnak SR, Motonaga KS, Skinner JR, Marcondes LD, Perry JC, Collins KK, Seslar SP, Cabrera M, Uzun O, Cannon BC, Aziz PF, Kubul P, Tanel RE, Valdes SO, Sanis I, Kertesz NJ, Malbonado J, Erickson C, Moore JP, Asaki H, Mill L, Alcedo M, Spector ZZ, Menon S, Shwyuyder M, Bradley Dj, Cohen M, Sanatani S. Life-threatening event risk in children with Wolff-Parkinson-White syndrome: a multi-center international study. JACC Clin Electrophysiol 2018;4:433–444.
4. Munger TM, Packer DL, Hammill SC, Feldman BJ, Bailey KR, Ballard Dj, Holmes DR, Gersh BJ. A population study of the natural history of Wolff-Parkinson-White syndrome. Circulation 1989;80:866–873.
5. Sharma ADY, Guiraudon R, Klein G; J G. Sensitivity and specificity of invasive and non-invasive testing for risk of sudden death in Wolff-Parkinson-White syndrome. J Am Coll Cardiol 1997;10:373–381.
6. Li CH, Hu YF, Lin YJ, Chang SL, Lo LW, Ta-Chuan T, Lee P-C, Huang S-Y, Sueari K, Tung NH, Tai C-T, Chao T-F, Chiang C-E, Chen S-A. The impact of age on the electrophysiological characteristics and different arrhythmia patterns in patients with Wolff-Parkinson-White syndrome. J Cardiovasc Electrophysiol 2011;22:274–279.
7. Page RL, Joglar JA, Caldwell MA, Callens H, Conti JB, Deal Bj, Estes NAM, Field ME, Goldberger ZD, Hammill SC, Indik JH, Lindsay BD, Olshansky B, Russo AM, Shen W-K, Tracy CM, Al-Khatib SM. 2015 ACC/AHA/HRS guideline for the management of adult patients with supraventricular tachycardia: a report of the American College of Cardiology/American Heart Association task force on clinical practice guidelines and the Heart Rhythm Society. J Am Coll Cardiol 2016;67:e27–e115.
8. Santinelli V, Radinovic A, Manguso F, Vicedomini G, Gulletta S, Paglino G, Mazzone P, Ciconte G, Sacchi S, Sala S, Pappone C. The natural history of asymptomatic ventricular pre-excitation a long-term prospective follow-up study of 184 asymptomatic children. J Am Coll Cardiol 2009;53:275–280.