

# Wolff-Parkinson-White as a bystander in a patient with aborted sudden cardiac death



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## Introduction

Sudden cardiac death (SCD) in young athletes is always dramatic, because unexpected, and is sometimes a challenge to investigate. The prevalence is estimated at 0.6 per 100,000 in the United States.<sup>1</sup> Coronary anomalies are the second most common cause of SCD in young athletes (16%) but are, unfortunately, difficult to recognize before a serious clinical event. Wolff-Parkinson-White (WPW) syndrome accounts for only 1.6% of all SCD.<sup>1</sup>

We present the case of a young athlete with aborted SCD and both WPW syndrome and an anomalous right coronary artery (RCA) origin.

## Case report

A 19-year-old student with no prior cardiac history presented with aborted SCD while playing hockey at his university arena. He was resuscitated by his teammates using an external automated cardiac defibrillator. He reported palpitations and chest discomfort before passing out.

He had no family history of SCD or syncope. Two years before, he had a normal resting electrocardiogram (ECG), negative exercise testing, and a normal Holter monitoring after complaining of chest pain and palpitations.

The physical examination was normal. The initial echocardiogram showed a structurally normal heart. The baseline ECG (Figure 1A) showed intermittent preexcitation with a positive delta wave in leads V<sub>1</sub>, DII, DIII, and aVF and negative delta wave in DI and AVL. This was consistent with a left anterolateral accessory pathway. Since the preexcitation was intermittent, the hypothesis that the WPW was responsible for his SCD had to be challenged. An electrophysiological study (EPS) was performed and confirmed the presence of an accessory pathway. The preexcitation was intermittent even on isoproterenol infusion, suggesting a

low-risk accessory pathway, since the anterograde refractory period was 320 ms (Figure 2A). Orthodromic tachycardia was induced on isoproterenol infusion with a tachycardia cycle length of 315 ms (Figure 2B).

Furthermore, during the isoproterenol infusion, antero-septal ST-segment elevation was documented (Figure 1B). The accessory pathway was successfully ablated without complication using the retrograde aortic approach. Because we already had arterial access for the ablation, we performed a coronary angiogram in the context of dynamic ECG changes during the isoproterenol infusion (supplemental material, available online). An abnormal RCA origin from the left sinus of Valsalva just above the left main artery was documented. Cardiac computed tomography (CT) confirmed a “slit-like” RCA coursing between the aortic root and the pulmonary trunk (Figure 3A). This was equivalent to a 70% stenosis at rest.

During cardiac surgery, the surgeon described a proximal aortic intramural course of the RCA treated by translocation of the ostium to the right sinus using an “unroofing technique” combined with an aortoplasty. Postoperative CT confirmed the complete correction (Figure 3B).

At 3 months follow-up, the patient did not report any recurrence of palpitation and had a normal ECG. He underwent a stress test myocardial perfusion scintigraphy with no evidence of residual ischemia. He resumed his normal activities, including hockey, with no events after 24 months follow-up.

## Discussion

The overall risk of SCD in WPW syndrome is estimated at 0.1% per year<sup>2</sup> in asymptomatic patients and 0.3% per year in symptomatic patients.<sup>3</sup> Ventricular fibrillation is generally secondary to atrial fibrillation, leading to extremely rapid ventricular response in the presence of an accessory pathway with critically short anterograde refractory period (usually <250 ms). EPS has a central role in assessing the electrophysiological properties of the accessory pathway. In our patient, a previous noninvasive investigation found no evidence of preexcitation and the stress test was negative despite complaints of palpitations and chest pain. Post

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**KEYWORDS** Sudden cardiac death; WPW syndrome; Anomalous origin of coronary arteries; Clinical electrophysiology; Investigation of sudden cardiac death (*Heart Rhythm Case Reports* 2016;2:399–403)

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## KEY TEACHING POINTS

- Aborted sudden cardiac death in young patients should trigger proper investigation to prevent recurrent events.
- Wolff-Parkinson-White syndrome could mask other causes of sudden cardiac death. Intermittent preexcitation is usually associated with a long pathway refractory period and should call into question the relationship with the malignant event.
- Cardiac arrest or chest pain during vigorous exercise in a young patient should mandate exclusion of an abnormal course of coronary arteries as a cause.

resuscitation, the ventricular preexcitation was intermittent and the echocardiogram showed no structural abnormality. We performed the EPS to confirm the presence of the accessory pathway and found it to be benign even on isoproterenol infusion. Because orthodromic tachycardia was inducible, the ablation was performed. With ST-T changes on isoproterenol infusion and history of aborted SCD during vigorous exercise, we completed the investigation with an angiogram to exclude an anomaly of the coronary arteries.

Intermittent loss of preexcitation at physiological heart rates has high specificity to identify patients at very low risk for SCD. However, Pauriah et al<sup>4</sup> found that baseline EPS missed 16% of patients at risk of life-threatening arrhythmias when isoproterenol infusion was performed. Moreover, Gemma et al<sup>5</sup> reported a patient with intermittent preexcitation and the shortest R-R interval in atrial fibrillation at 230 ms. In our patient, even on isoproterenol infusion, the accessory pathway anterograde refractory period remained long.

Both WPW and coronary anomalies are well-known causes of SCD. Their occurrence in the same patient is relatively novel. Yagita et al<sup>6</sup> published a case of WPW syndrome associated with an anomalous RCA origin from the left sinus responsible for anterolateral ischemia on exercise scintigraphy. It was suspected that a left coronary steal phenomenon occurred during exercise to explain the ischemic territory, since the RCA origin is just above the left main artery. In our patient, during isoproterenol infusion, the ST-segment elevation was anteroseptal in location, probably related to the same phenomenon. It is, however, unknown if orthodromic tachycardia in our patient could have favored ischemia in the presence of his coronary anomaly.

The prevalence of athletic field SCD has been estimated at 0.6 per 100,000 per year<sup>1</sup> in high school-

age athletes in the United States registry. Maron et al<sup>1</sup> reported 690 exertional cardiovascular-related SCDs in young athletes from 1980 to 2006. The main causes of death were hypertrophic cardiomyopathy (251/690; 36%) and coronary anomalies (119/690; 17%). Anomalous left main coronary artery from the right sinus of Valsalva was found in 65 patients and an anomalous RCA from the left sinus in 16 patients. In comparison, WPW syndrome was involved in only 11 of 690 patients (1.6%).

These coronary artery anomalies are difficult to diagnose before a catastrophic event. Indeed, Basso et al<sup>7</sup> reported 27 sudden deaths in young athletes identified at autopsy with left main coronary artery from the right sinus (n = 23) or RCA from the left sinus (n = 4). In this review, it was proposed in case of strong clinical suspicion and normal screening tests (resting ECG, stress test) to explore those anomalies with noninvasive tests such as transesophageal echocardiography focused on coronary artery anatomy, cardiac CT, or magnetic resonance imaging.

The optimal surgical management for these anomalies (bypass surgery, reimplantation, or unroofing techniques) remains controversial. Bypass surgery is associated with a higher rate of graft thrombosis<sup>8</sup> because of the competitive flow from the native vessels. This flow competition can be controlled by ligating the proximal portion of the bypassed vessel, but then the distal flow will rely on a conduit that might eventually suffer from attrition. Reimplantation surgery requires extensive dissection and does not address an intramural course. In the unroofing procedure, the ostium of the RCA is modified at the aorta by excising ("unroofing") the common wall located between the aorta and the anomalous coronary artery. This procedure is associated with excellent results. In a single center experience, Sharma et al<sup>9</sup> reported a low morbidity and mortality with no aortic valve incompetency in 75 patients. Two of them needed right internal mammary artery-to-RCA grafting owing to flow acceleration at the RCA ostium. All patients remained free of cardiac symptoms at a mean 18 months follow-up. There was 1 noncardiac death.

## Conclusion

This case report invites us to remain vigilant and critical when assessing an aborted SCD. Even in the presence of a WPW pattern on the resting ECG, a benign accessory pathway cannot lead to ventricular fibrillation and other differential diagnoses must be considered. It also reminds us that congenital coronary artery anomalies are the second most common cause of SCD in young athletes, and symptoms at exertion, if present, should be appropriately explored by transesophageal echocardiography, cardiac CT, coronary angiography, or magnetic resonance imaging if routine tests are negative.

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