

Congenital Atrioventricular Block and Wolff-Parkinson-White Syndrome

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We report the case of a patient followed since childhood for congenital complete atrioventricular block. At 28 years of age, atrioventricular conduction through an accessory pathway with long conduction times was detected. Periods of atrioventricular conduction alternated with periods of atrioventricular block. Sinus tachycardia and 1:1 exclusive conduction through the accessory pathway developed with increased sympathetic activity (exercise, isoproterenol infusion). We discuss the special features of this case.

Key words: *Cardiac block. Wolff-Parkinson-White Syndrome.*

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Bloqueo auriculoventricular congénito y síndrome de Wolff-Parkinson-White

Presentamos el caso de una paciente seguida desde la infancia por bloqueo auriculoventricular completo (BAVC) congénito. A los 28 años se detectó una conducción auriculoventricular (AV) a través de una vía accesoria con tiempos de conducción largos que alternaba con rachas de BAVC. Con el aumento de la actividad simpática (ejercicio, infusión de isoproterenol) se produjo una taquicardia sinusal y conducción 1:1 exclusiva a través de la vía accesoria. Se comentan las peculiaridades de este caso.

Palabras clave: *Bloqueo cardíaco. Síndrome de Wolff-Parkinson-White.*

INTRODUCTION

Congenital complete atrioventricular block (CAVB) occurs rarely, occurring in approximately 1 of every 20 000 newborns. The occurrence of atrioventricular (AV) accessory pathways with anterograde conduction (Wolff-Parkinson-White syndrome [WPW]) is estimated to occur in 1 out of every 1000 to 2000 people. Therefore, the association between congenital CAVB and WPW syndrome should be extremely infrequent, appearing in 1 of every 20 to 40 million births. In addition, it is possible that some cases would not be recognized because of the presence of normal cardiac frequency.

CLINICAL CASE

A patient was diagnosed with CAVB in our center at the age of 8 years. Patient tracings that the patient

brought with him showed a sinus rhythm of approximately 75 beats/minute, with CAVB and an escape rhythm of 55 beat/minute with a narrow QRS of normal morphology (Figure 1). On a Holter monitor performed when the patient was 21 years of age, CAVB could be seen throughout the entire recording, with a narrow escape rhythm of between 45 and 75 beats/minute. He lived several years without requiring revision and was referred to our center by his primary physician for the first time at the age of 28 years.

When he first visited our center the patient was asymptomatic. Sinus rhythm with CAVB and a narrow escape rhythm was observed on a ECG; it alternated with conducted sinus beats with a 200 ms PR interval and complex wide QRS with initial filling, positive in V1 and negative in III and VF, and suggestion of conduction by an inferior left AV accessory pathway (Figure 2). An echocardiogram showed the absence of structural cardiopathy.

In a 24-hour outpatient ECG recording, alternance between 2 rhythms was observed: during the periods of rest CAVB with escape of around 50 to 55 beats/minute with a narrow QRS and during sinus rhythm activity with 1:1 conduction and wide QRS. No pathological pauses or tachyarrhythmias were observed.

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Recibido el 30 de marzo de 2001.
Aceptado para su publicación el 18 de septiembre de 2001.

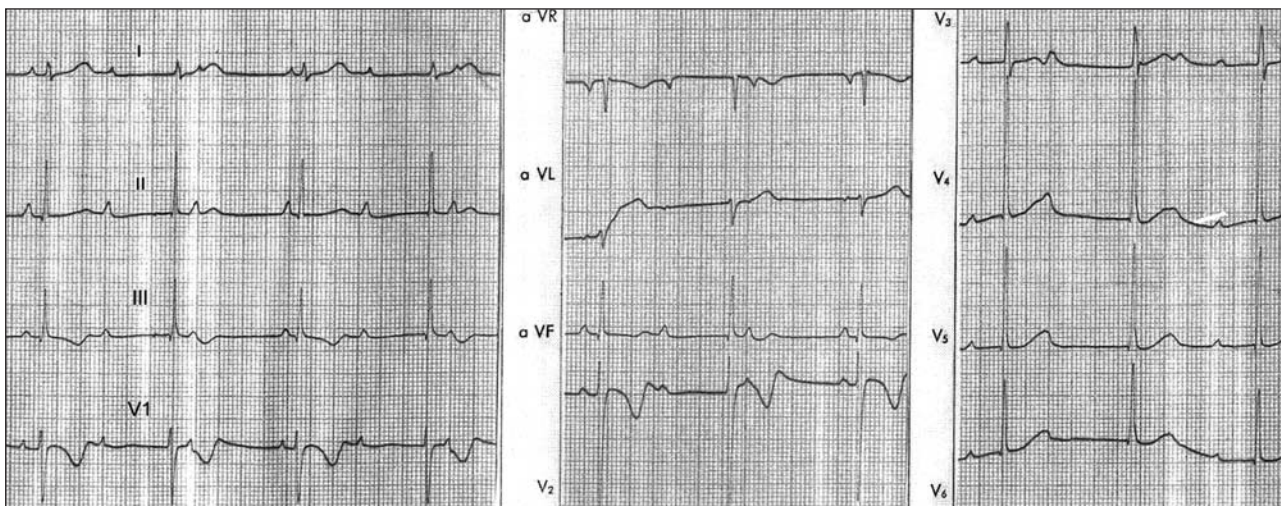


Fig. 1. ECG performed when the patient was 8 years of age. A complete atrioventricular block with an escape rhythm with narrow QRS can be seen.

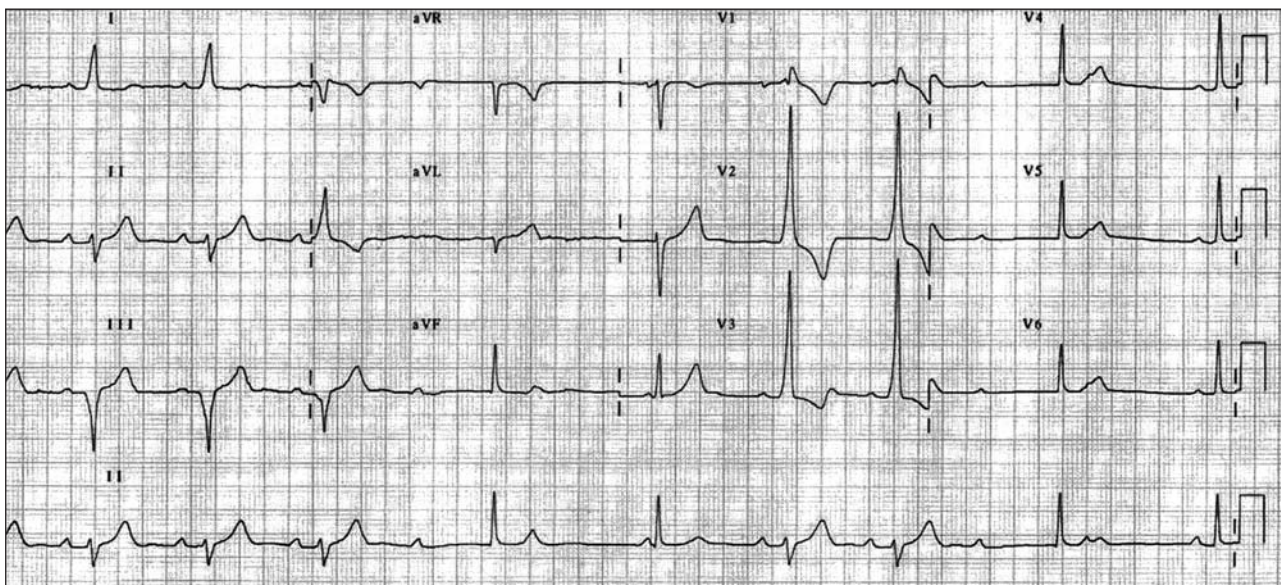


Fig. 2. ECG of the patient at 28 years of age. Conducted P-waves with a PR of 200 ms can be seen with a QRS morphology that suggests pre-excitation through an inferior left pathway alternating with blocked P-waves and a narrow escape rhythm.

Bruce protocol ergometry was performed, interrupted by rest at 9 minutes, that showed from the initiation of exercise a 1:1 conduction with a wide QRS up to 163 beats/minute (Figure 3), with slight lengthening of the PR interval (not visible on the image). During isoproterenol infusion, tachycardia and new conduction exclusively through the AV accessory pathway was observed, with maintenance of the PR interval (Figure 4).

DISCUSSION

The association between CAVB and the WPW syndrome is very infrequent. We have come across

only 3 cases in the literature that describe this association.¹⁻³ In all of them, the accessory pathway manifested at an earlier age than in the case we describe (in 2 cases it appeared in newborns and in the other at 7 years of age).

Although conduction by an accessory pathway is an abnormal occurrence and in many cases causes a pathological process, in our case it is, in fact, beneficial as it assures good AV conduction during exercise. Also, the patient did not have a history of a drop in atrial fibrillation. As the AV node is not functional, there is no possibility of orthodromic or antidromic tachycardia (although there are some exceptional cases described with CAVB and

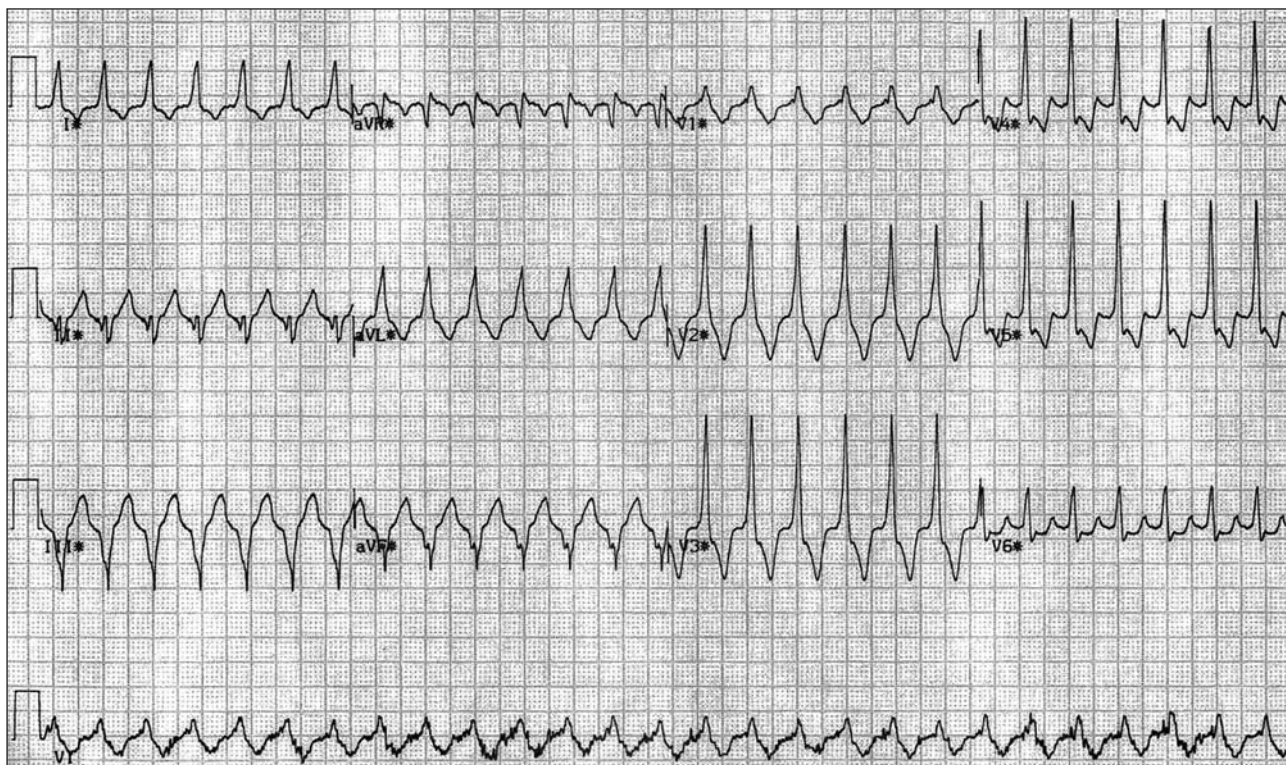


Fig. 3. ECG at the end of the stress test. Conduction 1:1 through the accessory pathway at 160 beats per minute. The P-waves are masked in the ST segment.

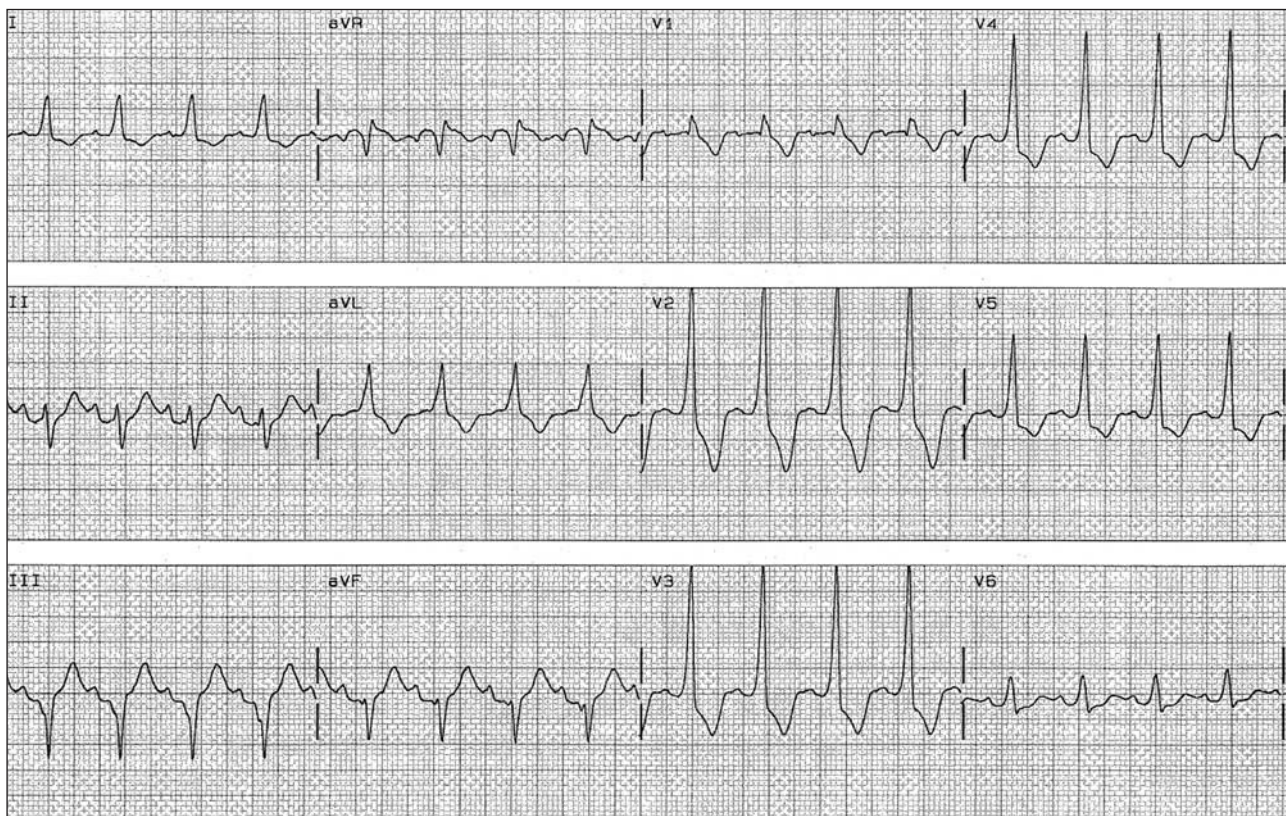


Fig. 4. ECG during isoproterenol infusion. Conduction 1:1 through the AV accessory pathway, with a PR interval similar to baseline.

retrograde AV conduction, which theoretically would not totally exclude the possibility of an antidromic tachycardia).

There are various points in this case that stand out: the lengthened PR interval, the not excessively wide conducted QRS complexes, and the late appearance of conduction through the accessory pathway. The QRS morphology is very suggestive of conduction by an accessory pathway, but the PR interval is slightly prolonged which is uncommon in a conventional WPW syndrome, in which the PR interval in fact tends to be shortened. This probably is due to a pathway with long conduction times, with poor baseline conductivity and properties of decreasing conduction.¹ Sympathetic stimulation (exercise, isoproterenol) would improve its conductivity,⁴ but the increase in cardiac frequency would manifest it decreasing conduction characteristics, slightly increasing the AV conduction time.

The QRS complexes conducted through the accessory pathway last less than 140 ms; given that they are completely pre-excited complexes, a greater width would be expected. Early penetration of the stimulus conducted through the pathway in the specific conduction system may explain this.

More difficult to explain is the appearance so late (in the third decade of life) of conduction by accessory pathway. It is logical to think that the histological basis (myocardial bridges between the atrium and the ventricle) was present from birth and that, therefore, the absence of conduction during these years may be due to the functional incapacity of the pathway to transmit the impulses. It could even be speculated that, given the great influence of sympathetic activity on this accessory pathway, both its prolonged latency and its actual functional situation could be linked to changes in the autonomous nervous system related to age.

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