BRIEF REPORTS

Congenital Atrioventricular Block and Wolff-Parkinson-White Syndrome

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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 at the age of 28 years.

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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 A V 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. 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 WPW syndrome).
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.

REFERENCES