A "multisite" atrioventricular block

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A 6-year-old child, operated for a perimembranous ventricular septal defect, underwent an electrophysiologic study for the presence of first degree atrioventricular (AV) block and bifascicular block with episodes of type 1 and type 2 second degree AV block. Electrophysiologic study showed a considerable infra-His conduction delay (HV interval 170 ms) and spontaneous phases of infra-His type 1 second degree AV block. During incremental atrial pacing supra-His type 1 second degree AV block and 2:1 infra-His AV block were simultaneously observed and this condition persisted unmodified despite the intravenous injection of atropine. The block distal to the His bundle was considered functional.

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Introduction

Atrioventricular (AV) and intraventricular conduction disorders are well-known complications of congenital heart disease surgery. A better understanding of the anatomy and localization of the conduction system in patients with congenital heart disease and the improved surgical techniques have been responsible, in the last years, for the rapid decline in surgical AV block from 25 to 2%[1-4]. In most patients, AV block manifests immediately after the operation. However, approximately 60% of the patients do not necessitate permanent pacemaker implantation because AV conduction normally returns within 9-14 days after surgery[3,5]. With regard to the remaining 40% of patients, the occurrence of late-onset AV block can range from 2 days to 25 years after surgical repair[1,6,7]. On rare occasions, these patients may develop bradyarrhythmias that have been hypothesized to result from lesions at various levels of the AV conduction system[8-10].

The following case is an example of such "suspected" multisite AV conduction system disease.

Case report

A.V., aged 1 year, underwent a surgical repair for a perimembranous ventricular septal defect with a Gore-Tex patch, complicated by a transient (4 days) complete heart block. His preoperative ECG was completely normal, without conduction disturbances.

Two years later, sporadic asymptomatic episodes of type 1 second degree AV block were diagnosed.

At the age of 6 years, he was referred to our department for complete clinical and instrumental evaluation. A recent standard ECG showed asymptomatic episodes of type 1 and 2:1 second degree AV block associated with first degree AV block (long PR interval 260 ms) and bifascicular block: complete right bundle branch block (QRS duration 160 ms) and left anterior hemiblock (frontal QRS axis -45°) (Fig. 1).

Physical examination revealed an asymptomatic young boy of average build who was in no apparent distress and had stable vital signs. Cardiac examination was not significant. His echocardiogram showed no residual shunt, with normal left ventricular dimensions and function.

ECG Holter confirmed the presence of episodes of type 1 and type 2 second degree AV block with a mean heart rate of 54 b/min (range 40 b/min at 05:05 a.m. and 82 b/min at 09:19 p.m.) and no pauses > 2 s nor significant ST/T abnormalities.

The treadmill stress test was stopped at the beginning of the second step of the Bruce protocol owing to the lack of compliance by the patient.

After informed consent was obtained, electrophysiologic study was performed in the post-absorptive state. Although complete electrophysiologic evaluation was performed, this report deals only with the

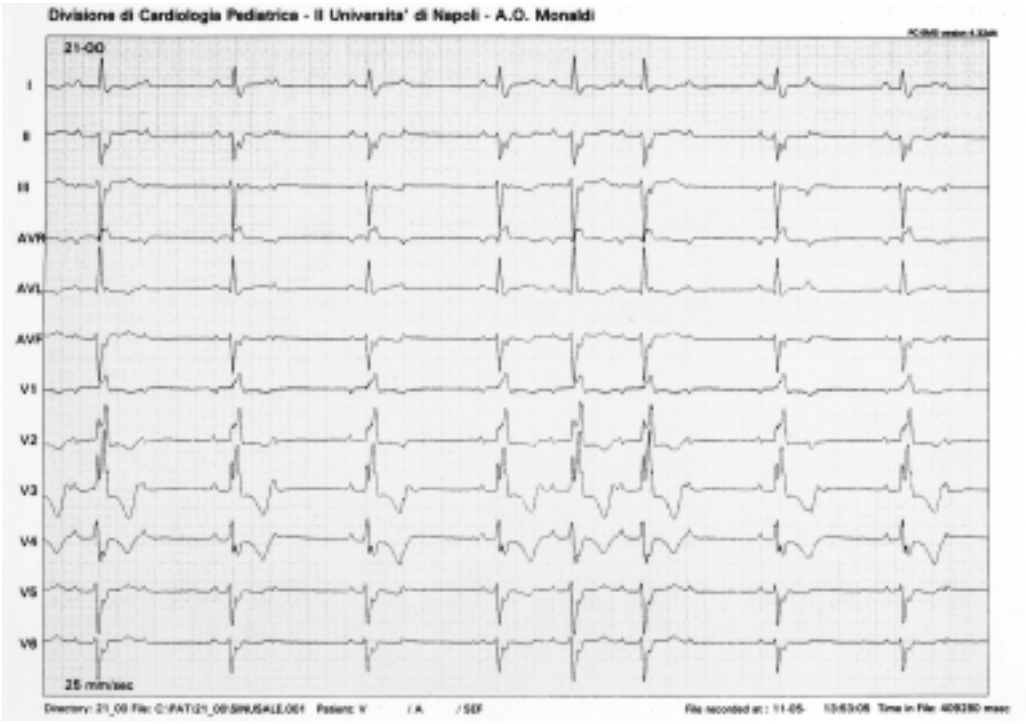

Figure 1. *Standard ECG: type 1 and 2:1 second degree atrioventricular block associated with first degree atrioventricular block and bifascicular block.*

results of the assessment of the conduction system. The electrophysiologic study was performed after mild sedation and using two 5F quadripolar catheters positioned in the high right atrium and His bundle region. The SA, AH, HV and VV intervals, as well as the conduction system refractory periods, were defined and measured as conventionally described.

The following data were obtained:
- normal sinus node function;
- normal supra-His conduction time (AH interval 76 ms);
- a considerable infra-His conduction delay (HV interval 170 ms), essentially located in the main His bundle fascicle, given that its potential preceded the right bundle one by a 150 ms interval and the right bundle deflection was anterogradely activated (20 ms) (Fig. 2). The validation of the His deflection was obtained both through atrial pacing and His bundle pacing;
- spontaneous phases of infra-His type 1 second degree AV block (Fig. 3);
- simultaneous recording of supra-His type 1 second degree AV block and infra-His type 2 second degree AV block during atrial pacing at a cycle length of 400 ms (Fig. 4), a condition that persisted unmodified despite the intravenous injection of 1 mg of atropine.

A prophylactic pacemaker was prescribed.

Discussion

Considerable concern has been expressed in recent years regarding the long-term prognosis of patients who develop various types of ventricular conduction disorders following open-heart surgery. Despite various pathological and clinical studies, significant controversy still exists regarding the precise localization of injury in many of these patients and their long-term prognosis. Electrophysiologic studies may help distinguish proximal from distal lesions. These abnormalities may be concealed and manifest only when the conduction system is stressed by pacing techniques. The usual response to incremental atrial pacing is the development of AV nodal Wenckebach periodicity (block proximal to the His bundle). A block distal to the His bundle induced by atrial pacing is a much less commonly encountered phenomenon, theoretically suggesting conduction abnormalities in the His-Purkinje system[11].

Fewer studies have analyzed pacing-induced 2:1 AV block during which two levels of block have been hypothesized to occur in two separate structures (AV node and His-Purkinje system)[8,9,12]. Otherwise, the presence of two-level conduction system abnormalities has been considered, for a long time, only conjectural, with very few electrophysiological evidences.

Our case clearly shows the possible coexistence of a proximal AH with distal HV conduction abnormality. However, in this patient, just as in other similar cases[11], the block distal to the His bundle has been considered to be functional.

Failure of impulse propagation at the AV node (during proximal AH Wenckebach periodicity) produces a long cycle length in the His-Purkinje system (long HH interval), with an associated long His-Purkinje refracto-

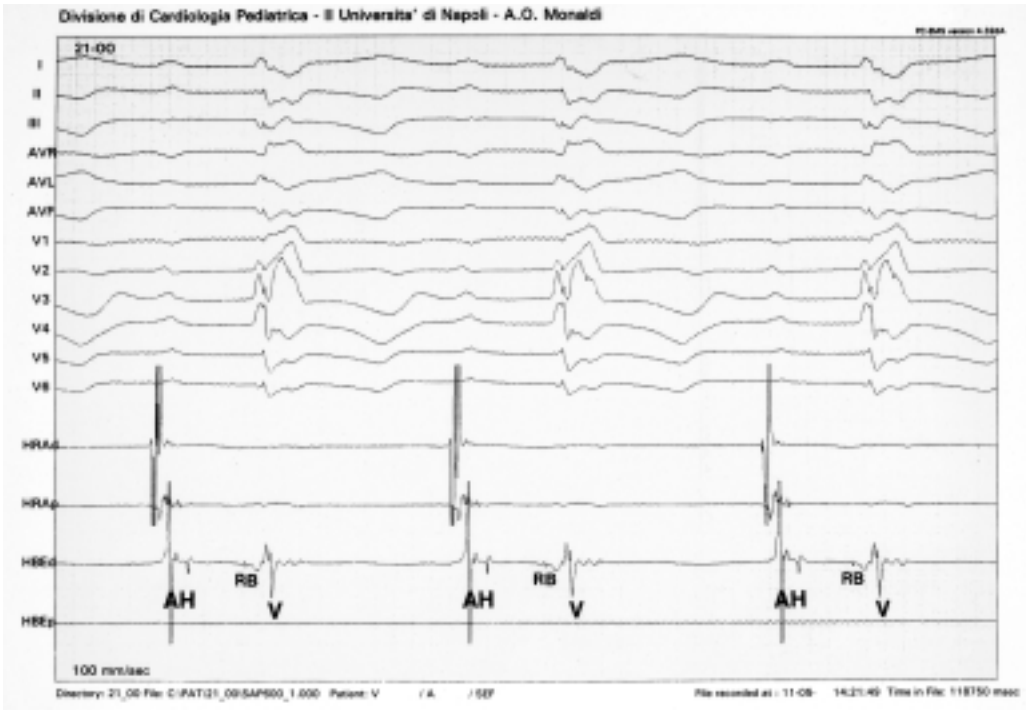

Figure 2. *Electrophysiologic study: infra-His conduction delay (HV interval 170 ms). A = atrial electrogram; H = His bundle electrogram; RB = right bundle electrogram; V = ventricular electrogram.*

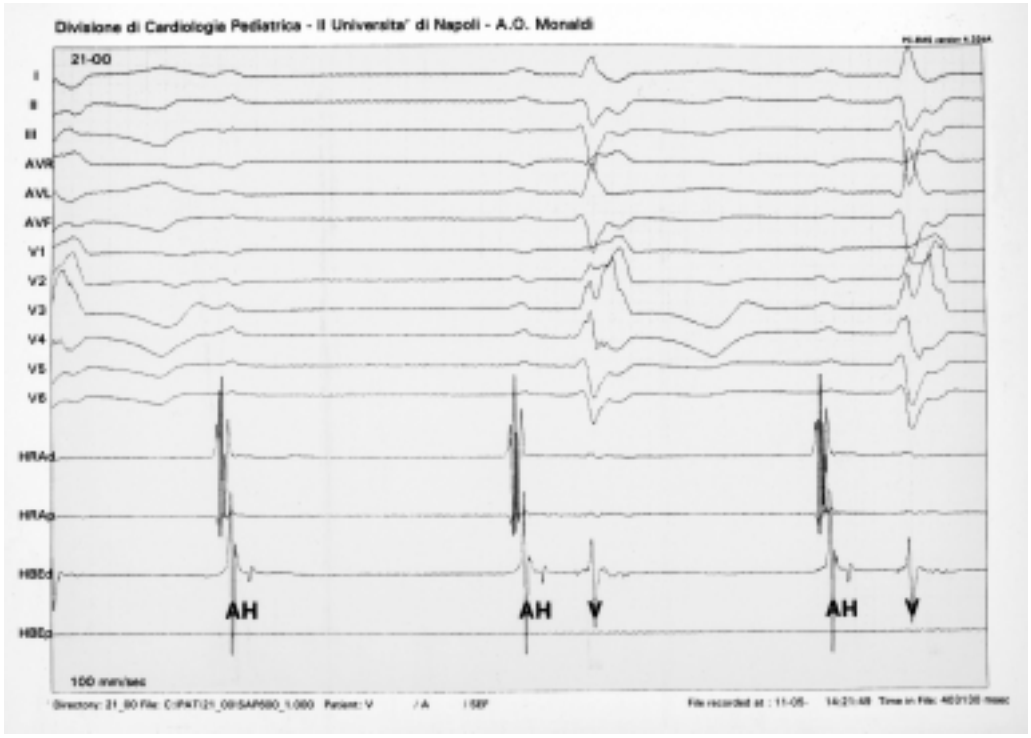

Figure 3. *Electrophysiologic study: spontaneous episodes of infra-His type 1 second degree atrioventricular block. A = atrial electrogram; H = His bundle electrogram; V = ventricular electrogram.*

ry period in the subsequent cycle (Ashman phenomenon). This prolongation of the His-Purkinje refractoriness results in a block distal to H of the second beat of the next Wenckebach sequence (short HH cycle) (Fig. 4).

This case study underscores how complex postoperative AV block genesis might result. Besides, it is also noteworthy that this disorder may be delayed and develop long after surgery. It is important to note that

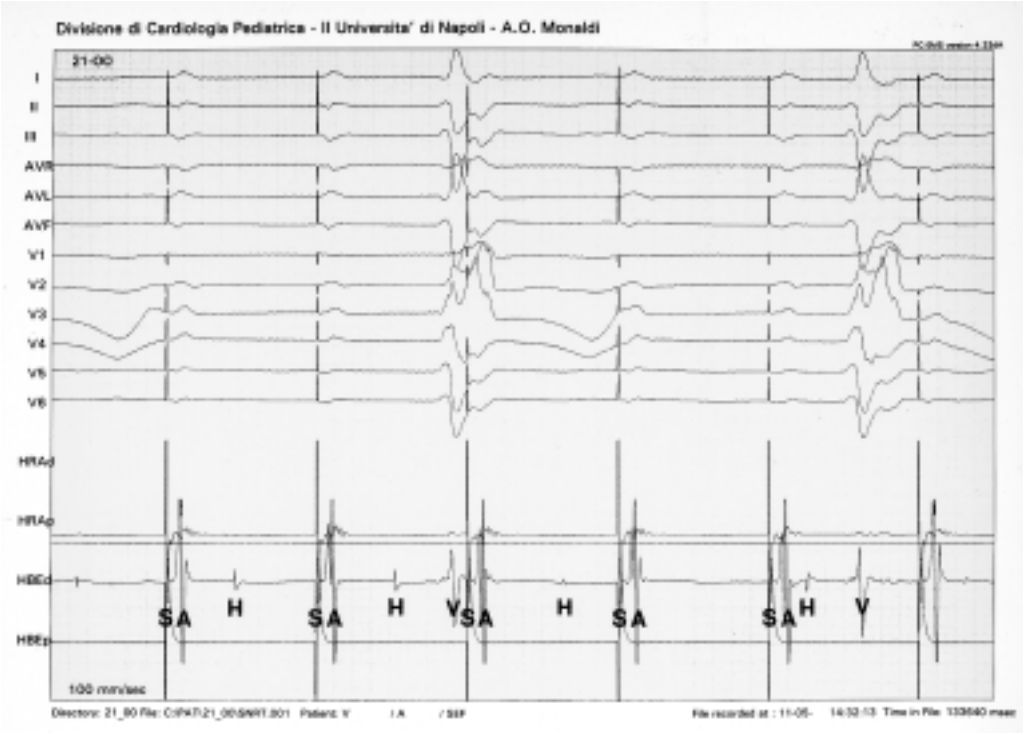

Figure 4. *Electrophysiologic study: simultaneous recording of supra-His type 1 second degree atrioventricular block and infra-His type 2 second degree atrioventricular block during atrial pacing at a cycle length of 400 ms. A = atrial electrogram; H = His bundle electrogram; S = spike; V = ventricular electrogram.*

even patients who seemingly recover from complete heart block early have the potential of presenting with delayed AV conduction abnormalities. This might be the case particularly for patients with transient complete heart block who regain AV conduction but have a residual intraventricular conduction abnormality. Thus, patients with transient postoperative complete heart block should be followed with standard ECG and ECG Holter for the potential development of late AV conduction abnormalities[3].

Controversy exists about the treatment of these patients, particularly infants, when implantation of an endocardial pacemaker would be technically difficult to implement[13].

Anyway, the finding of a third degree postoperative AV block that reverts to sinus rhythm with a residual bifascicular block is already a class II indication to pacemaker implantation both in national as well as in international guidelines[13,14]. Furthermore, in children no indication to pacing is now guided by the HV interval recording or by the response to pacing.

In the reported case, even in the presence of a "suspected" functional infra-His AV block, a prophylactic pacemaker implantation was prescribed for the demonstrated progression of the conduction system disease over time. No study has confirmed the effective value of this "aggressive" management in the prevention of sudden death and whether "functional" infra-His AV block has the same potential risk as "true" anatomic block.

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