

Intermittent atrio-ventricular block and ectopy in an infant following complex heart surgery: Occam's razor versus Hickam's dictum

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A 9-month-old male infant with double outlet left ventricle (DOLV), subpulmonary stenosis, and anomalous left coronary artery underwent a Rastelli type of biventricular repair at our institution. This entailed enlargement of the ventricular septal defect (VSD), left ventricle-to-aorta intraventricular VSD tunnel repair, and a right ventricle-to-pulmonary artery conduit. During the second postoperative week, he was noted to have frequent episodes of intermittent atrio-ventricular (A-V) block. In addition, during that same period, there was the emergence of sustained and nonsustained supraventricular tachycardia, as well as isolated supraventricular ectopy. He was hemodynamically stable without any inotropic support and his electrolytes were normal.

Based on the Telemetry/ECG provided (Figure 1), what is the diagnosis and management?

1 | COMMENTARY

Second- or third-degree A-V block is not uncommon after complex cardiac surgery in such infants. The persistence of Mobitz Type II second-degree A-V block beyond the 7th postoperative day should prompt careful monitoring. Progression to advanced second- or third-degree A-V block is a class I indication for permanent pacemaker implantation.¹ However, careful analysis of the ECG is warranted in order to accurately ascertain the etiology of any A-V conduction disturbance.

The initial telemetry tracing (Figure 1A) demonstrates a nonconducted sinus P wave (denoted by inverted red triangle; ▼) without any preceding PR or PP interval prolongation. This is ostensibly suggestive of Mobitz Type II second-degree A-V block.² The conducted sinus beats show a wide QRS morphology (100 ms; normal <80 ms in

an infant) with initial sharp intrinsicoid deflection and terminal slurring consistent with aberrancy. The second tracing (Figure 1B) shows premature wide-complex beats (denoted by arrows; ↓) with partially obscured nonconducted sinus P waves (denoted by inverted red triangle; ▼) occurring at its terminal portion. The QRS morphology of the premature beats is also slightly different from the conducted beats. Thus these represent either fascicular or junctional extra-systole with additional rate-dependent aberrancy.¹ The third tracing (Figure 1C) shows apparent premature atrial complexes or PACs (denoted by inverted purple triangles; ▼) followed by wide-complex beats/tachycardia beats (denoted by arrows; ↓) with a morphology similar to the previously noted premature QRS complexes. This is indicative of a conducted atrial ectopy/tachycardia. The fourth tracing (Figure 2) shows the combination of findings noted in Figure 1A and B.

To summarize, we ascertained an electrocardiographic triad of Mobitz Type II second-degree A-V block, fascicular/junctional extra-systole with aberrancy, and atrial tachycardia in this patient. It is possible that the above entities were isolated manifestations and mechanistically unrelated. An alternative unifying diagnosis is feasible based on the rationale enunciated subsequently.

Before arriving at a diagnosis in such complex scenarios, clinicians are often obligated to contemplate two competing philosophies in the medical decision-making process. The primal directive pertains to the well-known rules of diagnostic parsimony postulated by the principle of "Occam's razor." Antagonistic to this is the lesser known, nevertheless salient "Hickam's dictum." This is expounded by the aphorism *patients can have as many diseases as they damn (or darn) well please* attributed to eminent American physician John Hickam. A diagnosis of premature QRS complexes mechanistically unrelated

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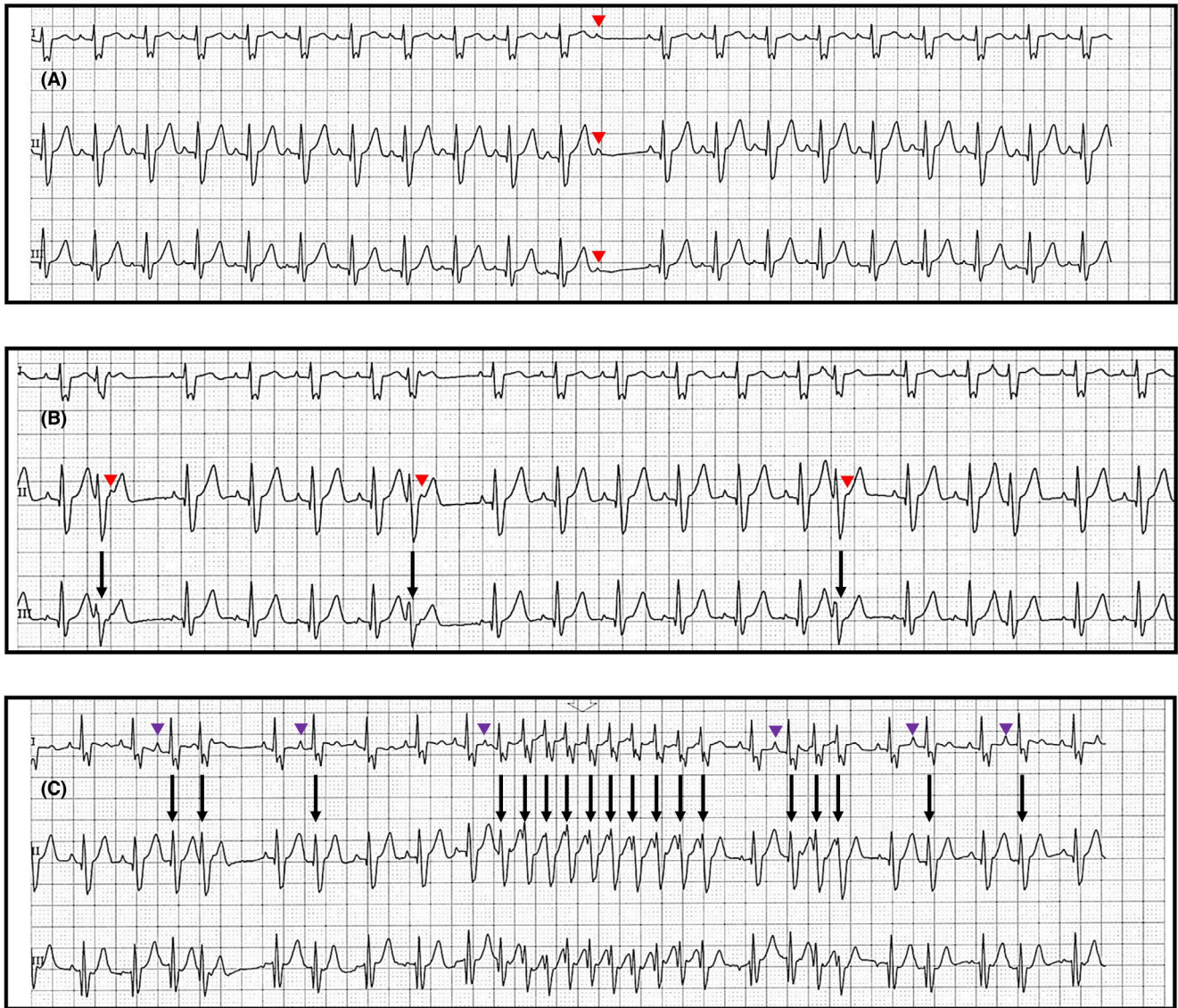


FIGURE 1 Telemetry strips of the patient. (A) The rhythm strip shows sinus rhythm with constant PR interval and aberrant conduction as well as a single nonconducted sinus beat (denoted by an inverted red triangle; ▼). (B) This rhythm strip shows premature wide QRS complexes (denoted by arrows; ↓) with partially obscured nonconducted sinus P waves (denoted by an inverted red triangle; ▼) occurring towards the end of the QRS. (C) This tracing shows frequent premature atrial complexes (denoted by an inverted purple triangle; ▼) followed by single or multiple beats of wide complex tachycardia similar in morphology to the one noted in Figure 1B

to concurrent atrial tachycardia or transient Mobitz Type II second-degree A-V block would follow the latter tenet.

However, in this case, the application of the principle of Occam's razor as explicated below lead to a different inference. A mechanistic "link" between premature QRS complexes and A-V block is conceivable by postulating the occurrence of critically timed "concealed" premature junctional/fascicular extra-systole as illustrated in Figure 2 (denoted by dotted arrows). An extra-systole originating within the proximal conduction system at a time when the distal conduction system is fully refractory will not produce any manifest QRS complex. The effect of such a premature beat can only be inferred if it conducts retrograde to the atrium or leads to "concealed" conduction within the A-V node. The latter scenario would render the A-V node either fully refractory causing A-V block (as in this case) or partially refractory leading to PR interval prolongation

for the next sinus beat.³ A succinct discussion of the possible diagnoses based on the ECG alone is warranted and has educational significance. To recapitulate, Hickam's dictum postulates that there are three co-existing diagnosis, namely Mobitz Type II second-degree A-V block, junctional extra-systole, and atrial tachycardia. Application of the Occam's razor allowed us to eliminate the possibility of Mobitz Type II second-degree AV block and instead attribute it to concealed junctional extra-systole. Thus the most rational diagnosis would be concealed and manifest junctional extra-systole with co-existing atrial tachycardia.

Although less likely, an alternative diagnosis to atrial tachycardia (Figure 1C) is conceivable if Occam's razor is refurbished again as proposed. The presumed PACs (denoted by inverted purple triangles; ▼) may surreptitiously represent junctional/fascicular extra-systole that only conduct retrograde to the atrium. The absence of associated

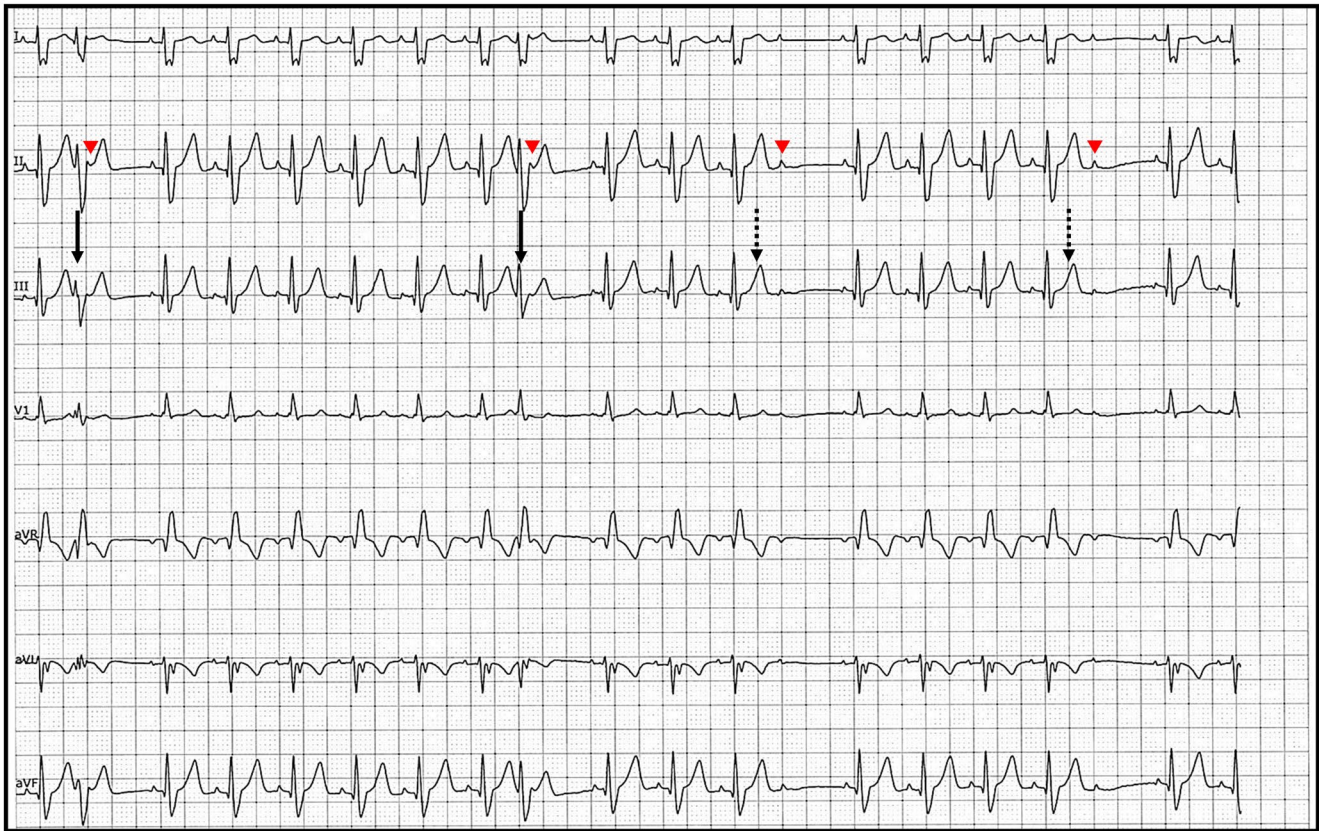


FIGURE 2 Telemetry strip shows premature wide QRS complexes (denoted by arrows; ↓). The nonconducted sinus P waves (denoted by an inverted red triangle; ▼) are easily discernable but become partially obscured if preceded by a premature QRS complex. A critically timed premature junctional extra-systole (denoted by dotted arrows) without a propagated QRS due to refractoriness of the distal conduction system is also presumed

manifest QRS complex is likely due to the distal antegrade functional block. This is likely followed by additional extra-systoles with manifest QRS complexes. The resulting junctional ectopic tachycardia could thus mimic a conducted atrial tachycardia. This hypothesis is speculative at its best and therefore left to the discretion of the aficionados of electrophysiology for thoughtful deliberation.

The surgical result was ascertained to be optimal based on echocardiographic evidence of normal biventricular size/function with patent outflow tracts and no A-V or semilunar valve regurgitation. Atenolol therapy was initiated, followed by complete suppression of all ectopy and any occurrences of “heart block.”

Pseudo A-V block due to concealed junctional extra-systole is previously reported in adults and is rarely noted in children.²⁻⁴ To the best of our knowledge, it is not described in infants. In our case, the cardiac surgery entailed anatomical repair adjacent to the conduction system. Postoperative junctional ectopic tachycardia is well-described in such circumstances and the findings reported here fall within that spectrum. In summary, meticulous ECG analysis played a pivotal role in accurately elucidating the etiology of this postoperative A-V conduction disturbance and obviated any extraneous therapeutic miscues.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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