Infant with 2:1 atrioventricular block after surgery for congenital heart disease: What's the mechanism?

Christopher L. Johnsrude, MD, MS

From the Division of Pediatric Cardiology, Department of Pediatrics, University of Louisville School of Medicine, Louisville, Kentucky, and Pediatric and Adult Congenital Heart Disease Arrhythmia Service, Norton Children's Hospital, Louisville, Kentucky.

Introduction

Surgery for congenital heart disease (CHD) results in injury to the atrioventricular node/His bundle (AVN/HB), causing postoperative AV block (AVB) in 0.7%–3%[1] and junctional ectopic tachycardia (JET) in up to 27%.[2,3] Primary risk factors for these arrhythmias include young age at surgery, specific surgical procedure, prolonged bypass/aortic cross-clamp times, and abnormal electrolytes.[2,3] Such arrhythmias can impose acute hemodynamic compromise, prolong hospitalization, and necessitate medical or surgical intervention.

Surface electrocardiograms (ECGs) facilitate diagnosis of these arrhythmias and guide clinical management. For most patients, JET responds to nonpharmacological measures or medications,[2,3] often resolving within a few days. Advanced AVB is managed with temporary pacing; a permanent pacemaker is recommended if present beyond postoperative day (POD) 7–10.[4] JET or AVB usually occur as isolated arrhythmias, but can manifest in the same patient.

Reported herein is a newborn with CHD whose early postoperative course was unremarkable until POD 6, when she developed new-onset 2:1 AVB and other arrhythmias, likely due to a seemingly rare mechanism, successfully treated with flecainide.

Case report

This full-term newborn underwent surgical repair of atrial and ventricular septal defects and aortic coarctation on day of life 2. Early postoperative rhythm was sinus with intact AV conduction and right bundle branch block (Figure 1A), common after VSD closure. Recovery was uneventful until POD 6, when continuous telemetry first recorded periods of 2:1 AVB (Figure 1B), prompting consideration for a permanent pacemaker.

Review of telemetry confirmed frequent isolated second-degree AVB and prolonged periods of 2:1 AVB, and other new arrhythmias. Frequent premature junctional (PJCs) or ventricular complexes (PVCs) were recorded, as were blocked premature atrial complexes (PACs) (Figure 1C). That evening, she developed sustained tachycardia triggered and terminated by PACs, with abrupt onset and stable rate (~180 beats/min), QRS morphologies identical to sinus beats, and no VA dissociation (Figure 2A and 2B). P waves during tachycardia were low-amplitude; 12-lead ECGs were not obtained.

One or multiple mechanisms could explain these arrhythmias, manifesting in the setting of evolving postoperative inflammation, resolving perioperative ischemia, or another condition; serum electrolytes were normal. Second-degree AVB could reflect interrupted conduction from injury to the AVN/HB, manifesting a bit later than usual. Late-coupled PACs could reflect abnormal atrial automaticity, blocking antegrade owing to the same AVN/HB injury. The sustained tachycardia suggested a reentrant mechanism, such as AVN or AV or atrial reentry. Rapid conduction during tachycardia suggested robust antegrade AV conduction, tough to square with intermittent AVB. The PJCs/PVCs might be caused by abnormal automaticity involving the AVN/HB, or sinus beats conducting in a 1:2 ("double-fire") fashion.[5,6] Of these possibilities, 2 mechanisms—HB extrasystoles (HBEs) and 1:2 AV conduction—might explain most of these arrhythmias, neither implicating intrinsic AVN/HB conduction disease.

Referral for a pacemaker was deferred and oral propranolol 0.5 mg/kg administered every 6 hours to treat tachycardia. After no effect on any of the above-mentioned arrhythmias for 36 hours, propranolol was exchanged for oral flecainide 1 mg/kg every 12 hours. Second-degree AVB and tachycardia did not recur after the first flecainide dose, and PACs and PJCs became rare, completely resolving within 24 hours. She was discharged home on POD 12 and continued on flecainide over 3 months of follow-up. Serial ECGs and 24-hour Holter monitors recorded no arrhythmias.

KEYWORDS AV block; Congenital heart surgery; His bundle extrasystoles; Infant; postoperative arrhythmia
(Heart Rhythm Case Reports 2022;8:27–30)

Funding Sources: The authors have no funding sources to disclose. Disclosures: The authors have no conflicts of interest to disclose. **Address reprint requests and correspondence:** Dr Christopher L. Johnsrude, Associate Professor, Division of Pediatric Cardiology, Department of Pediatrics, University of Louisville School of Medicine, Suite 113, KCPC, 571 S. Floyd St., Louisville, KY 40202. E-mail address: cljohn02@louisville.edu.

https://doi.org/10.1016/j.hrcr.2021.10.007

KEY TEACHING POINTS

- His bundle extrasystoles can manifest as diverse arrhythmias, including second-degree AV block, premature atrial and junctional complexes, and narrow complex tachycardia.
- Postoperative AV block following surgery for congenital heart disease can be due to "pseudo-block" from concealed His bundle extrasystoles (HBEs), including 2:1 AV block when HBEs occur in a bigeminal pattern.
- When evaluating a patient with an acute onset of diverse arrhythmias, consider that a single unifying mechanism is most likely responsible.

Discussion

This report describes a newborn with postoperative CHD developing advanced second-degree AVB, frequent blocked PACs, PJCs/PVCs, and narrow complex tachycardia, all starting on POD 6. Diverse pathophysiologies could be involved: intrinsic AVN/HB conduction disease, abnormal atrial and/or junctional automaticity, disruption of normal atrial inputs to the AVN, and sustained reentrant supraventricular (SVT) or atrial tachycardia. Review of telemetry tracings suggested His bundle extrasystoles might well explain the majority of these arrhythmias. Resolution of all arrhythmias followed initiation of oral flecainide, obviating recommendation for pacemaker implantation. Although possible that these diverse arrhythmias resolved spontaneously, the timing coincident to beginning flecainide was quite compelling.

Diverse bradyarrhythmias or tachyarrhythmias from injury to the compact AVN, His bundle, or AVN artery after CHD surgery typically manifest soon after rewarming, and are transient or permanent. AVB initially presenting after the first several days is uncommon, likely reflecting ongoing postoperative inflammation or scarring. AVB is often isolated, but some patients also have junctional, atrial, or ventricular tachyarrhythmias. If AVB is present beyond POD 7–10, a permanent pacemaker is recommended.[4]

Postoperative JET also usually presents in the first hours postbypass. ECGs show tachycardia warming up and cooling down, consistent with abnormal phase 4 automaticity of the compact AVN or His bundle. Tachycardia rates are influenced by endogenous and exogenous catecholamines and serum electrolytes. AV dissociation during JET occurs when junctional rates exceed sinus rates and retrograde block prevents resetting of the sinus node. Intact antegrade AV conduction is confirmed when properly timed sinus beats or atrial overdrive pacing above the JET rate entrains ventricular rhythm; these findings are absent when JET is accompanied by AVB. JET often responds to lower core temperature, sedation, enhanced preload, treating anemia and hypokalemia, and antiarrhythmic agents.[2,3] JET may be overdrive-suppressed but not triggered or terminated by PACs, burst atrial pacing, or direct current cardioversion. JET manifesting very early after surgery and responding to treatment usually resolves within a few days. JET first presenting several days after surgery is comparatively rare, and can last longer and be more difficult to suppress.

An uncommon AVN-related arrhythmia involves 1:2 or "double-fire" AV conduction of sinus/atrial impulses. ECG patterns typically show uninterrupted background sinus rhythm with "downstream" AV conduction alternating between the fast (FP) and slow pathways (SP),[6] which on ECG appears as "PJCs." If FP-SP conduction occurs repetitively, frequent PJCs cause ventricular rates to be twice that of sinus rhythm. At other times, sinus rhythm may conduct with abruptly changing PR intervals, when antegrade conduction is intermittently blocked in the FP owing to retrograde concealment from the SP.[5] In our patient's case, the appearance of frequent PJCs/PVCs could potentially be explained by this mechanism. However, our patient's sinus rhythm was interrupted when PJCs/PVCs appeared, and abruptly changing PR intervals were not recorded. Our patient's sustained SVT was clearly not due to consecutive FP-SP conduction, as the sinus P waves disappeared during tachycardia (Figure 2). SVT was induced by a PAC followed by a long AV time, potentially implicating AVN reentrant tachycardia as the tachycardia mechanism. Although one might expect AVN reentrant tachycardia to occur frequently in patients with ECG evidence of dual AVN physiology like double-fire conduction, this combination is quite rare[5] and not reported in pediatric patients. In addition, frequent blocked PACs and sustained periods of 2:1 AVB of sinus beats have not been reported in patients with double-fire AV conduction. Finally, double-fire AV conduction rarely responds to antiarrhythmic agents, instead requiring treatment with catheter ablation of the slow pathway to the AV node.[5]

Review of ECG recordings suggest most of our patient's arrhythmias could reflect manifest or concealed HBEs; confirmatory electrophysiology testing was not performed. HBEs result from abnormal focal automaticity called afterdepolarizations or triggered activity,[7] pathophysiology very different than common examples of postoperative AVB and JET. Clinical manifestations of atrial and ventricular afterdepolarizations have been reported in patients with long QT syndrome, digitalis toxicity, and other conditions.[7,8] The mechanism of HBEs was postulated in 1947 by Langendorf and Mehlman,[9] who deduced HBEs simulating first- and second-degree AVB and blocked PACs. HBEs were later confirmed by Rosen and colleagues[10] using intracardiac HB recordings. Whereas Nasrallah and colleagues[11] reported HBEs in 2 children with postoperative CHD aged 3 and 6 years old, most reports of HBEs involve adults.[12–14] Triggered activity does not respond to conventional treatments for phase 4 JET, but can be initiated and

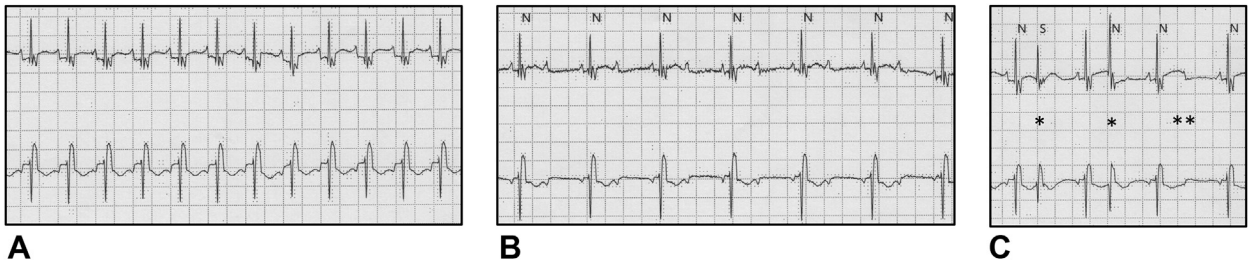

Figure 1 Postoperative telemetry recordings: **A:** Sinus rhythm with normal PR intervals and bundle branch block pattern. **B:** Sinus with 2:1 AV block. **C:** Sinus rhythm interrupted by frequent premature junctional complexes (*) and blocked premature atrial complexes (**).

terminated by premature extrasystoles or pacing, and suppressed with class I antiarrhythmic agents, calcium channel blockers, and adenosine.[7] The mechanism of our patient's recurrent narrow complex tachycardia remains enigmatic, as the telemetry recordings alone do not discern repetitive HBEs from more common forms of SVT such as AVN or AV reentrant tachycardia.

ECG diagnosis of His bundle extrasystoles

HBEs have been called "the great masquerader" because of myriad ECG manifestations.[13] Indeed, it is this diverse behavior and Occam's razor that facilitate diagnosis of HBEs by surface ECG alone.

First, recall that ECGs record summed atrial and ventricular depolarizations (P waves and QRS complexes, respectively), not specialized tissues like the sinus and AV nodes and His-Purkinje fibers. Second, HB afterdepolarizations can exhibit a fairly stable coupling interval related to the preceding HB depolarization, unlike varying R-R intervals seen with abnormal phase 4 automaticity of typical postoperative JET. Finally, clinical manifestations of HBEs depend on the frequency and timing of afterdepolarizations relative to prevailing cardiac rhythms, and on interactions with adjacent "upstream" compact AVN and atrial myocardium and "downstream" His-Purkinje system and ventricular myocardium.

Once initiated, an HBE conducting both upstream and downstream appears as a PJC coupled with a PAC. When the QRS complex is similar but not identical to sinus beats, aberrant conduction or more distal origin in a proximal bundle branch is suspected.[15] The P wave from the HBE may precede or follow the QRS complex, depending on propagation times to reach atrial and ventricular myocardium. The first PJC in Figure 1C was clearly followed by a PAC, and that reset the next sinus beat (Figure 3A).

HBE conduction will be blocked upstream or downstream when the impulse encounters adjacent structures still refractory from the preceding depolarization. We see that our patient had blocked PACs (Figure 1C) despite being only minimally premature, when a PAC would normally conduct downstream unless there was significant AVN/HB conduction disease. This finding could be explained by an HBE occurring when downstream conduction encountered ventricular refractoriness (hence no QRS), and retrograde atrial depolarization was affected by VA decrement in the AV node. In this way, an HBE conducting upstream but blocking downstream appears as a blocked "PAC" (Figure 3B). Late PACs bearing similarity to sinus P waves may reflect fusion of atrial depolarization from the HBE and subsequent sinus impulse (Figure 3B). An HBE blocking upstream but conducting downstream would appear as an isolated PJC (Figure 3C).

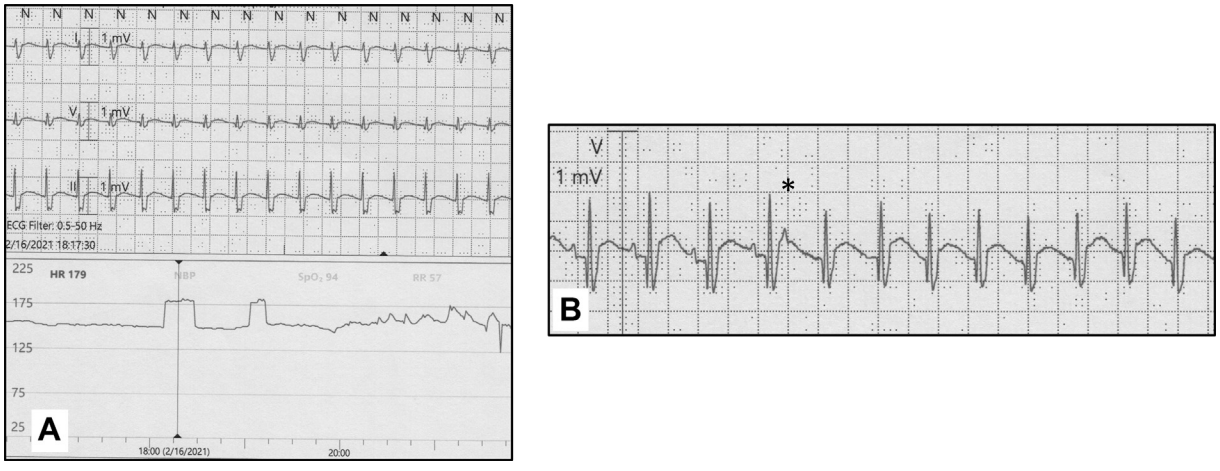

Figure 2 **A:** Sustained tachycardia with QRS complexes similar to sinus rhythm, abrupt onset and termination, and stable rate ~180 beats per minute. **B:** Premature atrial complex (*) induces tachycardia, sinus P waves are not readily apparent, and there was no clear VA dissociation.

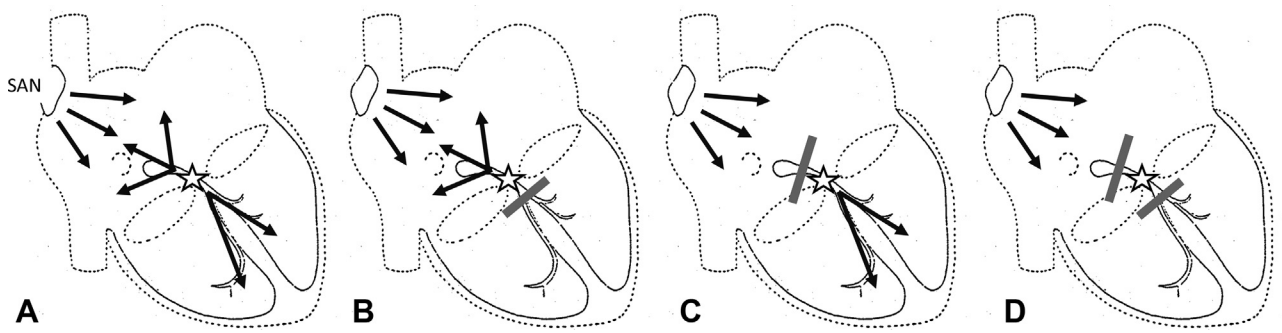

Figure 3 Background sinus rhythm and His bundle extrasystoles (HBEs). **A:** During sinus rhythm, an HBE conducts retrograde to the atrium and antegrade to the ventricles. **B:** HBE conducts retrograde to atria but blocks antegrade to the ventricles, causing a blocked premature atrial complex. **C:** HBE conducts to ventricles but blocks retrograde in atrioventricular node, resulting in an isolated premature junctional complex. D: HBE blocks in both retrograde and antegrade directions, not affecting sinus node automaticity yet blocking conduction of a sinus impulse to the ventricles. Star indicates His bundle extrasystole. SAN = sinoatrial node.

The occurrence of second-degree AVB in our patient (Figure 1B) causing the initial concern could be explained by pseudo-block from "concealed" HBEs. HBEs are concealed if the afterdepolarization occurs when both adjacent upstream and downstream structures are still refractory (Figure 3D). Blocked upstream conduction of the HBE impulse prevents depolarization of the atrial myocardium (hence no PAC) or resetting the sinus node. The HBE also initiates local refractoriness, which in turn prevents conduction of the subsequent sinus impulse through the HB, causing pseudo-AVB. The sinus impulse following the blocked beat does not encounter another HBE, and so conducts normally. Whereas our patient's isolated second-degree AVB was consistent with Mobitz II, HBEs can also appear as Mobitz I.[12] In addition, our patient's telemetry also recorded prolonged periods of 2:1 AVB, which could be due to concealed HBEs occurring in a bigeminal pattern.

Conclusion

This report describes 2:1 AVB in a young infant occurring 6 days after CHD surgery, raising consideration for implantation of a pacemaker. Given the simultaneous occurrence of numerous other arrhythmias, a unifying mechanism that could explain most of these arrhythmias was that of HBEs, with concealed HBEs causing pseudo-AVB and manifest HBEs causing PJCs/PVCs and blocked PACs, and possibly runs of narrow complex tachycardia. All arrhythmias resolved after initiation of flecainide therapy, and a pacemaker was not implanted. The patient has been arrhythmia-free on flecainide for over 3 months postoperatively. We suspect this arrhythmia arose from postoperative changes, and we anticipate trialing a "drug holiday" in the coming months. Although HBEs seem rare in clinical practice, some authors suggest they are likely more common, but are not often investigated or diagnosed if only manifesting as isolated PACs, PJCs, or PVCs.[14]

References

1. Lin A, Mahle WT, Frias PA, et al. Early and delayed atrioventricular conduction block after routine surgery for congenital heart disease. J Thorac Cardiovasc Surg 2009;140:158–160.
2. Moak JP, Arias P, Kaltman JR, et al. Postoperative junctional ectopic tachycardia: risk factors for occurrence in the modern surgical era. Pacing Clin Electrophysiol 2013;36:1156–1168.
3. Entenmann A, Michel M, Herberg U, et al. Management of postoperative junctional ectopic tachycardia in pediatric patients: a survey of 30 centers in Germany, Austria, and Switzerland. Eur J Pediatr 2017;176:1217–1226x.
4. Epstein AE, DiMarco JP, Ellenbogen KA, et al. 2012 ACCF/AHA/HRS Focused Update Incorporated Into the ACCF/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities. J Am Coll Cardiol 2013;61:e6–e75.
5. Wang NC. Dual atrioventricular nodal nonreentrant tachycardia: a systematic review. Pacing Clin Electrophysiol 2011;34:1671–1681.
6. Liu CF, Ip JE, Lin AC, Lerman BB. Mechanistic heterogeneity of junctional ectopic tachycardia in adults. Pacing Clin Electrophysiol 2013;36:e7–e10.
7. Clusin WT. Calcium and cardiac arrhythmias: DADs, EADs, and alternans. Crit Rev Clin Lab Sci 2003;40:337–375.
8. Wit AL. Afterdepolarizations and triggered activity as a mechanism for clinical arrhythmias. Pacing Clin Electrophysiol 2018;41:883–896.
9. Langendorf R, Mehlman FS. Blocked (nonconducted) A-V nodal premature systoles initiating first and second degree A-V block. Am Heart J 1947;34:500–506.
10. Rosen KM, Rahimtoola SH, Gunnar RM. Pseudo A-V block secondary to premature non-propagated His bundle depolarization. Documentation by His bundle electrocardiography. Circulation 1970;42:367–373.
11. Nasrallah AT, Gillette PC, Mullins CE, et al. Concealed His bundle extrasystoles in congenital heart disease. Am J Cardiol 1975;35:288–292.
12. Fisch C, Zipes DP, McHenry PL. Electrocardiographic manifestations of concealed junctional ectopic impulses. Circulation 1976;53:217–223.
13. Ameen A, Dharawat A, Khan A, et al. His Bundle extrasystoles revisited: the great electrocardiographic masquerader. Pacing Clin Electrophysiol 2010;34:e56–e59.
14. Jimenez-Diaz J, Lez-Ferre JJG, Heredia GU, Moreno-Planas J. Mobitz type II second-degree atrioventricular block with narrow QRS and junctional extrasystoles: what is the mechanism? J Cardiovasc Electrophysiol 2014;25:447–449.
15. Jazayeri M-R. Role of concealed conduction and allied phenomena in the genesis, maintenance, and termination of cardiac arrhythmias. Pacing Clin Electrophysiol 2019;42:779–804.