# Tachyarrhythmias arising from the conduction system in pediatric patients with complete heart block

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## Introduction

Tachyarrhythmias arising from components of the conduction system are well described and include entities such as atrioventricular (AV) nodal reciprocating tachycardia, junctional ectopic tachycardia, and fascicular ventricular tachycardias. There have been rare reports of such tachyarrhythmias arising in the setting of preexisting complete heart block (CHB).[1] We describe 2 cases of pediatric patients with congenital heart disease (CHD) and remote postoperative CHB presenting with arrhythmias arising from the conduction system.

## Case report
### Case 1

A 12-year-old female patient with a history of D-transposition of the great arteries/ventricular septal defect underwent arterial switch operation as an infant. This was complicated by CHB without a reliable junctional escape, for which she received an epicardial dual-chamber pacemaker at 1 week of life. Her atrial lead fractured 1 year postimplant and her device was reprogrammed to ventricular demand pacing. She was followed for typical pacemaker management without any complaints of or documented arrhythmias. At age 12 years, she presented to the emergency department with complaints of headache and tingling in her extremities. Her heart rate was noted to be 151 beats per minute, and electrocardiogram (ECG) showed a wide complex tachycardia with a right bundle branch block morphology and superior QRS axis with ventriculoatrial (VA) dissociation (Figure 1A), but she was alert, oriented, and normotensive. Device interrogation confirmed onset of the arrhythmia 4 hours prior. She otherwise had no known history of tachyarrhythmias, and her

only medication was losartan for mild ventricular dysfunction. She was admitted to the cardiac intensive care unit, where she was given 2 1 mg/kg boluses of lidocaine, with no change in her presenting rhythm. Magnesium was administered to optimize an incidental finding of mild hypomagnesemia (1.8 mg/dL), which transiently converted her rhythm to her baseline ventricular paced rhythm. Given the morphology and axis of the tachycardia, there was suspicion for fascicular ventricular tachycardia (VT); thus 2 doses of intravenous verapamil (5 mg each) were administered with conversion to ventricular paced rhythm. She was transitioned to oral verapamil with sustained rhythm control. An echocardiogram demonstrated mild left ventricular (LV) dilation and dysfunction, unchanged from her prior echocardiograms.

In the setting of the suspected diagnosis of idiopathic fascicular VT, based on the morphology of the presenting rhythm and the response to verapamil, the patient was taken to the electrophysiology (EP) laboratory. At baseline, the patient was in her ventricular paced rhythm with VA dissociation. Attempts were made to induce tachycardia with atrial pacing, but there was no conduction antegrade via pacing from the right atrium or from a catheter in the distal coronary sinus. The ablation catheter was then placed in the left ventricle via a retrograde approach and single ventricular extrastimulus testing was performed. At 400/320 ms, her clinical tachycardia was induced, with 12-lead correlation to her ECG in tachycardia, with VA dissociation and a V-V interval of 406 ms and atrial cycle length of 553 ms. No His bundle electrogram was present. The LV anterior and posterior fascicles were mapped, to assess for fascicular potentials and for local ventricular activation timing. Activation mapping in the region of the posterior mid LV septum demonstrated earliest signals in the region of the anticipated course of the posterior fascicle. Detailed mapping was performed in this region, and ultimately a diastolic potential was identified with timing ∼60 ms ahead of surface QRS. Multiple short radiofrequency (RF) applications in this region resulted in no change in rhythm, but there was poor catheter contact while in tachycardia. Tachycardia was pace-terminated, and the region of earliest diastolic potential was then targeted while in her baseline ventricular paced rhythm, which afforded better catheter stability. RF energy applied in this location resulted in spontaneous initiation of her clinical

## KEY TEACHING POINTS

- Postoperative permanent complete heart block does not preclude the possibility of having concomitant tachyarrhythmias arising from the distal conduction system.

- The bundle of His and atrioventricular node are not involved in the reentrant circuit of fascicular ventricular tachycardia; thus it is possible to observe this tachyarrhythmia in patients with complete heart block.

- Junctional ectopic tachycardia may be observed in patients with permanent complete heart block, the presence of which allows for a more aggressive ablation strategy.

VT and termination during the lesion. A line of ablation lesions was then completed across the mapped course of the left posterior fascicle, with no further inducible VT despite aggressive ventricular stimulation (S3 protocol down to ventricular effective refractory period at 400/230/230 ms) (Figure 1B and 1C). The patient was discharged following the procedure and electively returned to the device lab for placement of a new atrial lead to restore AV synchrony. The patient is now back to her normal activity level and her ECG demonstrates appropriate atrial-sensed, ventricular-paced rhythm, and she has not had recurrent VT in a year of follow-up.

### Case 2

A 7-year-old male patient with a single-inlet, double-outlet right ventricle, undetermined ventricular looping {S,X,L} with pulmonary atresia underwent staged single ventricle palliation to a fenestrated extracardiac Fontan circulation at age 3. The tricuspid valve (TV) was repaired at the time of Fontan in the setting of severe tricuspid regurgitation emanating from an anterior cleft using sutures and an annuloplasty ring; the procedure was complicated by the development of postoperative CHB. The patient had no underlying junctional escape rhythm and a permanent epicardial dual-chamber pacemaker was placed. Four years after this surgery, he was brought back to the operating room in the setting of severe TV regurgitation and moderate right ventricular systolic dysfunction for TV repair and upgrade to multisite pacing, with unremarkable intraoperative course and reassuring postoperative echocardiogram demonstrating a well-functioning TV with trivial regurgitation and no stenosis. On postoperative day 0, however, the patient developed junctional ectopic tachycardia (JET) to 170 beats per minute. He initially responded to sedation, cooling, and intravenous procainamide, allowing for atrial pacing and transient improvement in hemodynamics (Figure 2A). However, he

subsequently developed recurrent JET and procainamide was replaced with amiodarone. Despite aggressive antiarrhythmic therapy and escalating vasoactive infusions, the patient continued to demonstrate tenuous hemodynamics attributed to a combination of tachycardia and loss of AV synchrony, and ultimately required cannulation to VA extracorporeal membrane oxygenation.

Despite maximal medical management, the patient was unable to separate from VA extracorporeal membrane oxygenation. Given that the patient already had CHB, the decision was made to bring the patient to the EP lab for attempted ablation of the junctional focus in the hopes of restoring paced AV synchrony to allow for decannulation. Upon arrival to the EP lab, the patient remained in accelerated junctional rhythm/JET, with heart rates consistently >160 beats per minute. Femoral venous and arterial access were obtained and a transbaffle puncture was performed to access the atrium. Intracardiac tracings demonstrated VA dissociation with a slow atrial rhythm. Mapping the AV groove from both a transbaffle and a retrograde aortic approach (Figure 2B) demonstrated earliest ventricular activation in a right posterior location (between 5 and 6 o'clock on the tricuspid annulus) near a gap in the partial annuloplasty ring. Transient arrhythmia termination related to a mechanical bump occurred in this region, but JET resumed within a minute. Precision mapping was performed at this location, ultimately identifying an area of local ventricular activation 40 ms pre-QRS (Figure 2C). RF ablation performed at this site resulted in abrupt termination of the arrhythmia within 1 second of coming on ablation, at which point he reverted to his A-paced V-paced rhythm. Following ablation, there was no recurrence of accelerated junctional rhythm/JET despite 2 epinephrine boluses, and the underlying rhythm was now CHB with ventricular escape rate in the 30s–40s. Over the subsequent days, intravenous antiarrhythmics were weaned off and he was transitioned to daily oral amiodarone. The patient had no further recurrence of JET for the remainder of his hospital course.

### Discussion

In this case series, we describe 2 children with CHD who, despite longstanding CHB, presented with tachyarrhythmias arising from the cardiac conduction system: fascicular VT and JET.

### Fascicular VT

Idiopathic fascicular VT represents 10%–15% of VT related to the LV in structurally normal hearts and was first described by Cohen and colleagues[2] and Zipes and colleagues.[3] While fascicular VT may arise from the anterior or posterior fascicle, the most common type (90%) arises from the posterior fascicle[4] and is characterized by a relatively narrow ventricular rhythm with a right bundle branch block morphology with a superior/leftward axis, thought due to reentry with exit near the left posterior fascicle.[5] Although there are reports of fascicular VT in patients with minor CHD (atrial and
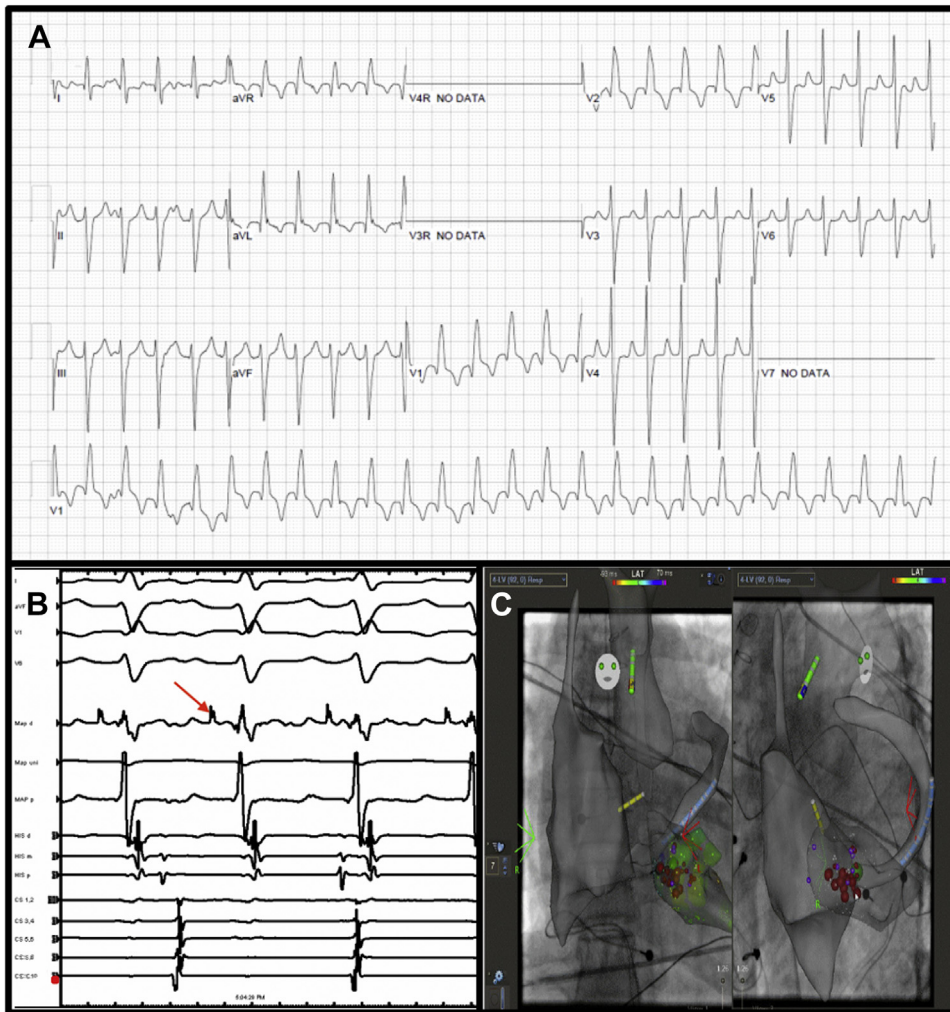
**Figure 1** **A:** Twelve-lead electrocardiogram with right bundle branch block morphology with a superior QRS with ventriculoatrial dissociation. **B:** Signal at site of successful ablation with the presence of diastolic potentials (*red arrow*). **C:** A 3-D electroanatomic map with right and left anterior oblique views showing site of ablation along the posterior fascicle.

ventricular septal defects, mitral valve prolapse),[4,6] to the best of our knowledge this represents the first report of fascicular VT in a patient with CHB. The mechanism of fascicular VT is assumed to be macroreentry within the Purkinje network in most patients.[7] It is believed that this reentry tachycardia stems from abnormal Purkinje fibers because of these specialized conduction tissues' dependence on the slow conduction of calcium in partial depolarization.[8] This is the basis for targeted therapy of fascicular VT with calcium channel blockers such as verapamil. Since the reentry circuit is entirely confined to the fascicles/Purkinje system, it is reasonable to assume that the bundle of His and AV node are not involved in this circuit and that such a tachycardia would be possible in a patient with CHB. Additionally, in this patient, preexisting CHB allowed for aggressive ablation along the posterior fascicle without the consequences of left bundle branch block secondary to ablation high along the fascicle.

## Junctional ectopic tachycardia

JET is an automatic tachyarrhythmia arising from the AV node and His bundle area and is most frequently seen in the postoperative pediatric population following corrective surgery for CHD, especially those surgeries involving manipulation or repair close to the region of the AV node. Decades ago, postoperative JET in children carried a high mortality,[9] and historically, His bundle ablation with pacemaker implantation had been used as an aggressive measure in refractory patients.[10] However, advances in the management of JET (including more careful manipulation intraoperatively near the AV node), controlled hypothermia, sedation, and intravenous antiarrhythmic therapies to decrease the junctional rate and allow restoration of AV synchrony with temporary pacemakers have largely supplanted such aggressive and irreversible interventions.[11,12]

Though it is commonly noted in the postoperative setting that JET and acute AV block coexist—likely owing to the

**Figure 2**    **A:** Telemetry strip demonstrating accelerated junctional rhythm. **B:** Right and left anterior fluoroscopic views demonstrating catheter approach retrograde through the aorta. **C:** Signal at site of success with high-frequency presystolic potentials, ∼40 ms pre–surface QRS (*red arrow*).

mechanism of both the automaticity and the injury being due to trauma, stretch, local edema, or ischemia in this region[13]—this case highlights the complexity of the AV node and proximal his bundle electrogram—such that even in the presence of longstanding CHB there is still the potential for arrhythmogenesis.[14] His bundle automaticity secondary to such changes may also be responsible for the JET that is seen in this setting and must be kept on the differential as a potential mechanism, but clinically cannot be differentiated from automaticity arising from the node itself.[13]

Unique to this case, the presence of preexisting permanent CHB allowed for a more aggressive invasive strategy of RF ablation after failure of optimized noninvasive management. RF ablation was performed in this patient without concern for inducing heart block, which has an incidence as high as 20% in some studies.[15]

## Conclusion

In this case report, we present 2 pediatric patients with repaired CHD and longstanding postoperative heart block and ventricular pacemaker dependency who presented with tachyarrhythmias arising from the distal conduction system. Invasive electrophysiologic study and ablation do not carry the same potential consequences as they could in a patient with intact AV nodal conduction, and may make the risk-benefit assessment of an invasive strategy favorable in unresponsive arrhythmias.

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