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Complete congenital heart block: A case of multilevel block.

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Introduction

Congenital heart block (CHB) has long been associated with maternal anti-SSA/Ro antibodies. Injury to the conduction system is presumed to be a consequence of the transplacental passage of maternal IgG autoantibodies into the fetal circulation.[1] Within the heart, a spectrum of phenotypes has been described, including structural defects, functional abnormalities, and electrical disturbances such as heart block. Third-degree atrioventricular (AV) block is typically irreversible, has a mortality rate approaching 30%, and requires pacemaker placement in up to 66% of cases.[2,3]

Autopsies of cardiac specimens from individuals with CHB have demonstrated the absence or partial absence of internodal and interatrial pathways.[4] Disruption of the structure of Bachmann's bundle, the major interatrial pathway, can result in interatrial dissociation in which each atrium beats independently.[5] Under this circumstance, the right atrium (RA) is under the control of the sinus node, while the left atrium (LA) is under the control of an ectopic pacemaker. We describe a newborn with complete CHB in the setting of maternal anti-SSA/Ro antibodies in whom interatrial dissociation was recognized after placement of a dual-chamber epicardial pacemaker system with bipolar LA and left ventricular (LV) pacing leads.

Case report

The patient was a newborn ex 35-week male infant born to a mother with Sjögren syndrome and anti-SSA/Ro antibodies with a prenatal and confirmed postnatal diagnosis of complete CHB. At approximately 18 weeks gestation, the fetus was noted to have CHB with accompanying AV valve insufficiency and biventricular dysfunction. The mother received a prolonged course of oral steroids as well as 2 doses of intravenous immunoglobulin with qualitative improvement in valvar and ventricular function. The fetal heart rate was unchanged by maternal therapies, however, and ranged from 45 to 55 beats per minute (bpm) throughout gestation. As the pregnancy progressed, the fetus developed an increasing pulmonary valve gradient. The patient was delivered at 35 weeks via planned cesarean section. Shortly after birth, heart rates ranged from 53 to 58 bpm. Umbilical lines were placed, and an infusion of epinephrine was initiated at 0.05 mcg/kg/min, which subsequently increased the heart rate to the low 60s.

The initial 12-lead electrocardiogram (ECG) demonstrated complete AV block and a junctional escape rhythm. The QTc was prolonged at 793 msec. The atrial rate was 117 bpm and the ventricular rate was 56 bpm. Careful inspection of the P-wave morphology revealed a narrow P wave (duration 40 msec) with absent negative terminal forces in V1, suggesting absent LA activation. Initial postnatal echocardiogram demonstrated thickened, doming pulmonic valve leaflets with moderate pulmonary valve stenosis, moderate pulmonary valve insufficiency, and normal biventricular systolic function. A 24 hour Holter monitor on day of life 1 demonstrated complete heart block with junctional escape rhythm, frequent isolated premature ventricular complexes, 1 ventricular couplet, and an average ventricular rate of 56 bpm. Epinephrine was initially titrated to maintain the heart rate greater than 60 bpm, and was later transitioned to isoproterenol owing to increasing frequency of ventricular ectopy, which subsequently improved.

On day of life 3, the patient was taken to the operating room for an uncomplicated placement of a Medtronic dual-chamber pacemaker (Medtronic, Minneapolis, MN) with bipolar LA and bipolar LV pacing leads. The pacemaker was initially placed in DDD mode with a rate range of 80–180 bpm and an AV interval of 120 msec. An initial postoperative ECG demonstrated an atrial-sensed ventricular-paced rhythm (93 bpm), 100% ventricular capture, and what appeared to be an independent atrial rhythm. The non-sensed P waves occurred at a rate of 112–120 bpm. Increasing the gain of the ECG 2 times revealed an additional smaller electrical signal (P wave), occurring 120 msec prior to every QRS, indicated by the arrows in Figure 1. Pacemaker interrogation revealed an atrial capture threshold of 0.5 volts at 0.5 msec

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KEY TEACHING POINTS

- Infants of anti-SSA/Ro antibody–positive mothers demonstrate a spectrum of pathologic conduction abnormalities.
- Interatrial dissociation is possible in infants born to mothers with anti-SSA/Ro antibodies.
- In interatrial dissociation, placement of a left-sided pacing system allows for coordination of left atrial and left ventricular contraction.
- It appears that there is a natural progression of the left atrial impulse to become more unstable with time.

and ventricular capture threshold of 0.5 volts at 0.5 msec; the atrial electrogram measured 1.4 mV and the ventricular electrogram measured 4 mV.

At 2 months of age, the patient was noted to have increasing pulmonary valve stenosis with a peak pulmonary valve gradient approaching 60 mm Hg. He was subsequently referred for balloon pulmonary valvuloplasty, which occurred without complication at 3 months of age. An ECG at 2 months of age demonstrated the normalization of his QTc interval to 420 ms. At a subsequent outpatient follow-up visit at 5 months of age, pacemaker interrogation revealed stable capture and sensing thresholds. A rhythm strip obtained during pacemaker interrogation demonstrated initial variability in heart rate. By the end of the strip, loss of spontaneous LA activity was observed, suggesting instability of the LA focus. Despite this, non-sensed P waves continued to march through at a rate of 150 bpm (Figure 2). A second rhythm strip obtained after the lower rate limit of the pacemaker was temporarily programmed to 160 bpm demonstrated P waves marching through at a rate of 150 bpm (Figure 3). At that visit, the permanent lower rate limit was programmed to 80 bpm. Remote device interrogation 1 week later demonstrated 100% AV-paced rhythm at a rate of 80 bpm, further suggesting instability of the LA focus, as there was no pacing above the lower rate limit.

Discussion

In the presented patient, it became clear that as management progressed there was evidence for multilevel conduction block, at both the AV nodal and interatrial level. This electrophysiologic observation has not been previously reported in a newborn infant presenting at birth with complete CHB. Initially, our patient was thought to have isolated complete CHB with normal sinus node function. Only after placement of the left-sided dual-chamber pacing system did it become clear that the native P wave from the RA was in complete electrical isolation from the LA. Interatrial conduction block was confirmed after numerous pacemaker interrogations demonstrated the inability to either track or overdrive capture the native RA P wave. Interatrial conduction block was suggested by the following observations:

(1) No electrical resetting of spontaneous LA activity by RA activity, despite the spontaneous RA rate being faster than LA activity (Figures 1 and 2).
(2) Dissociation of RA P-wave activity during an LA-sensed ventricular-paced rhythm during DDD pacing (Figures 1 and 2).
(3) Failure of conduction from the LA to the RA at a pacing rate faster than the RA rate (Figure 3).
(4) Following development of spontaneous absence of LA activity, there was no RA-to-LA conduction (Figure 2).

Transplacental passage of maternal anti-SSA/Ro antibodies to the fetal circulation is associated with an increased risk of fetal CHB. More than 60% of affected children require lifelong pacemakers before entering adulthood owing to varying degrees of AV block or sinus bradycardia.[3] A unique aspect of CHB is that it arises from an injury during fetal development, as it has only been reported once in the maternal heart.[6] The pathogenesis is thought to result from 2 processes: apoptosis and calcium channel blockade by the anti-SSA/Ro antibodies.[3] The unique interplay between 3 variables—the maternal autoantibodies, the in utero environment, and a genetic predisposition—has been theorized to cause the wide variety of phenotypes observed.[6] A number of electrophysiologic disturbances have been described, including various degrees of AV block and sinus bradycardia. Autopsy studies of children born with CHB have demonstrated a variety of pathologic abnormalities in the conduction system. These have included a lack of connection between the atria and the AV node, complete absence of the AV node, lack of connection between a demonstrated AV node and the His bundle, and discontinuity of the His bundle itself.[4]

Bachmann's bundle, also known as the interatrial bundle, is a muscular bundle containing parallel aligned myocardial strands connecting the right and left atrium. It is considered to be the main pathway of interatrial conduction.[5] Disruption of the bundle's structure causes interatrial conduction block, which is associated with development of various atrial tachyarrhythmias as well as electromechanical dysfunction of the LA.[7–10] Bachmann's bundle contains cells similar to those found within the SA node and the AV node and shares electrophysiologic properties of both Purkinje and atrial fibers.[5]

In an autopsy case series by James et al,[4] the cardiac conduction system of 3 siblings with CHB were examined. Each of the 3 hearts studied had similar abnormalities in the sinus node, internodal and interatrial pathways, and AV node. In 1 of the cases, there was almost total absence of the interatrial and internodal pathways, whereas the other 2 specimens demonstrated lesser degrees of interatrial pathway damage.

Interatrial conduction block may promote spontaneous atrial arrhythmias (atrial flutter or fibrillation) and impair hemodynamics by compromising the ability to facilitate proper timing of LA-to-LV AV contraction necessary to maximize

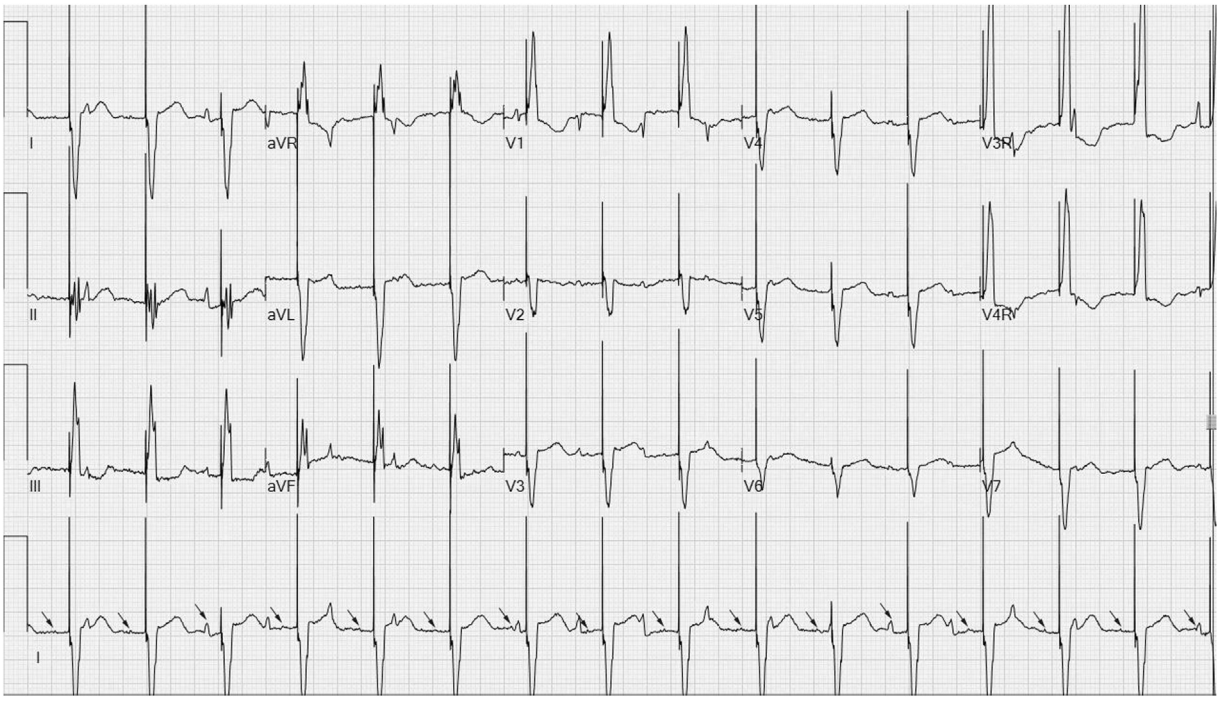

Figure 1 Initial electrocardiogram post dual-chamber pacemaker placement at 2× gain demonstrating atrial-sensed ventricular-paced rhythm at 93 beats per minute (bpm), noncaptured P waves at a rate of 112–120 bpm, and low-amplitude left atrial impulse prior to every QRS complex. (Arrows indicate left atrial impulse.)

cardiac output.[7–10] Placement of an LV epicardial pacing lead is considered to be the optimal choice for the ventricular lead in neonates with CHB, as it is less likely to contribute to pacing-induced LV dyssynchrony and possible development of pacing-induced cardiomyopathy.[10,11] Placement of a left-sided pacing system also allows for coordination of LA and LV contraction, typically not necessary when RA-to-LA conduction is normal. If a right-sided pacing system (RA lead) had been placed in our patient, RA to either right or left ventricular pacing would perpetuate left interatrial dissociation from left ventricular contraction, decreasing cardiac output from the lack of contribution of coordinated LA-to-LV systole.

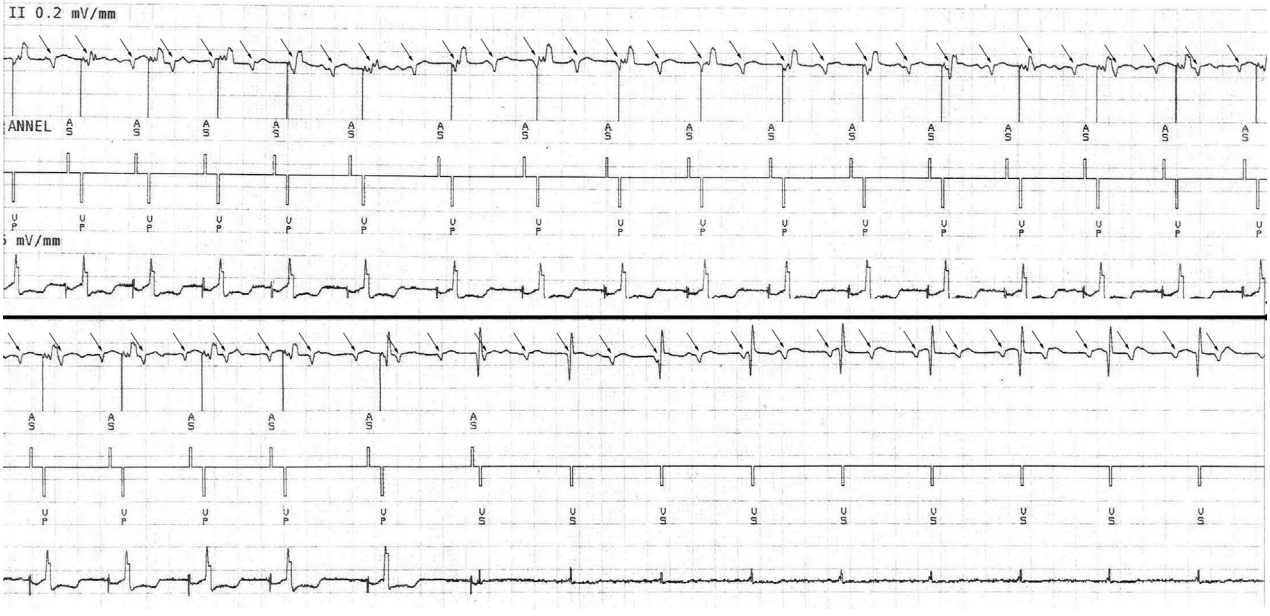

Figure 2 Rhythm strip from pacemaker interrogation at 5-month outpatient visit. Strip demonstrates initial variability in heart rate (top row) and by the end of the strip (lower row) there is loss of spontaneous left atrial activity and accelerated junctional rhythm. Nonconducted P waves march through the entirety of the strip at a rate of 150 beats per minute. (Arrows indicate right atrial impulse.)

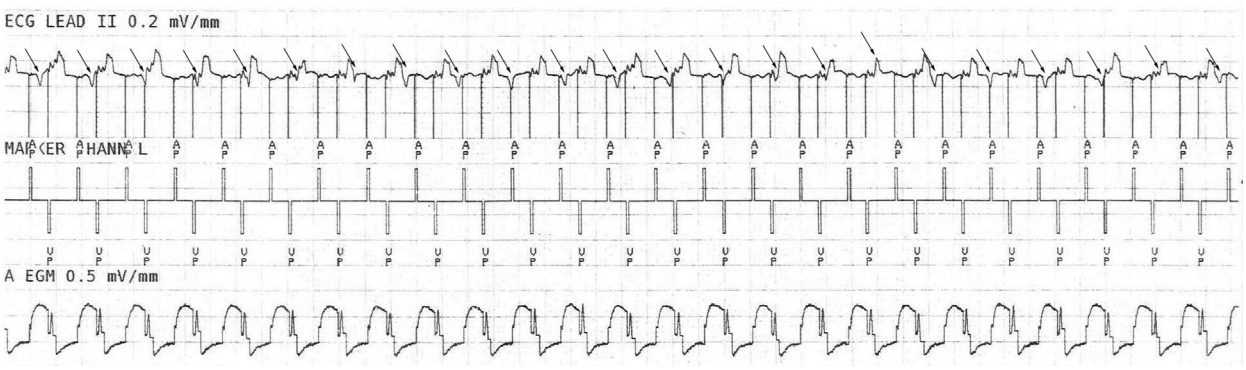

Figure 3 Rhythm strip from pacemaker interrogation at 5-month outpatient visit, during which the lower rate of the pacemaker was temporarily programmed at 160 beats per minute (bpm), demonstrating P waves continuing to march through at a rate of 150 bpm. Left atrial capture observed after atrial pacing spike. (Arrows indicate right atrial impulse.)

Furthermore, the natural progression of the LA impulse to become more unstable and without the presence of an LA lead to allow for continuous LA pacing will promote periods of LA asystole, known to be detrimental by predisposing to LA thrombosis, as occurs during atrial fibrillation.

The prevalence of interatrial block in this population is currently unknown but is likely rare or perhaps underdiagnosed. Ultimately, it is clinically important to assess this finding in similar patients to ensure optimal hemodynamics through coordination of LA and LV contraction.

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