Infrahisian Heart Block in Pediatric Patient with Congenitally Corrected Transposition of the Great Vessels and Ebstein's Anomaly.

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Introduction

- SLL-Transposition of the Great Arteries and Ebstein's Anomaly are two congenital heart defects known to have complications with the conduction system
- SLL-TGA being predisposed to conduction delay due to the compact AV node being distant from the bundle of his
- Ebstein's anomaly is conducive development of accessory pathways and atrial arrhythmias
- SLL-TGA with Ebstein's Anomaly is a rare, but defined abnormality which poses unique risk for arrhythmias and conduction system abnormalities

Clinical Presentation and Management

5 year old with congenitally corrected transposition of the great arteries and Ebstein's anomaly develops infrahisian heart block, requiring placement of an epicardial pacemaker.

Patient had been followed conservatively with low normal systemic right ventricular systolic function and mild tricuspid regurgitation. He had an episode of SVT, likely AVRT as an infant, with no other sustained tachyarrhythmias in early childhood. His medications included digoxin, metoprolol and enalapril. Patient presented as infant to the emergency room with hypoglycemia and unresponsiveness thought to be due to accidental extra ingestion of beta blocker. 12 lead ECG revealed sinus bradycardia to 109 bpm with first degree AVB. His beta blockade was stopped and digoxin decreased. He was observed in the cardiac ICU and noted to have sinus bradycardia, and occasional Wenckebach. He was discharged on digoxin only, 10 mcg/kg/day. In follow-up, he was noted to have low heart rates to 60 bpm per parental auscultation. Digoxin was decreased by half with subsequent heart rates 100-110 bpm. Clinic ECGs demonstrated sinus bradycardia in 90-100 bpm over the following months. 8 months later, an ECG was obtained for a telehealth visit. This revealed second degree AV block (Wenckebach), which was a change from baseline. The patient had no overt symptoms of significant heart block by clinical history. His digoxin was discontinued. A Holter was obtained which demonstrated second degree AV block, mostly Mobitz type 1 with signs of infrahisian heart block and rare third degree heart block. A stress ECG was obtained which confirmed demonstrated infrahisian heart block. Repeat Holter monitor after a sedated CMR revealed 3rd degree heart block. An epicardial dual chamber biventricular pacemaker was implanted. Due to his history of mild systemic ventricular dysfunction which improved over time, a loose pulmonary artery band was placed at the same time in hopes of improving his interventricular hemodynamics given his risk of systemic right ventricular systolic failure over time. At his first pacemaker check, his underlying conduction was 3rd degree heart block (sinus rate of 105 bpm with narrow complex ventricular escape of 48 bpm).

ECG Progression

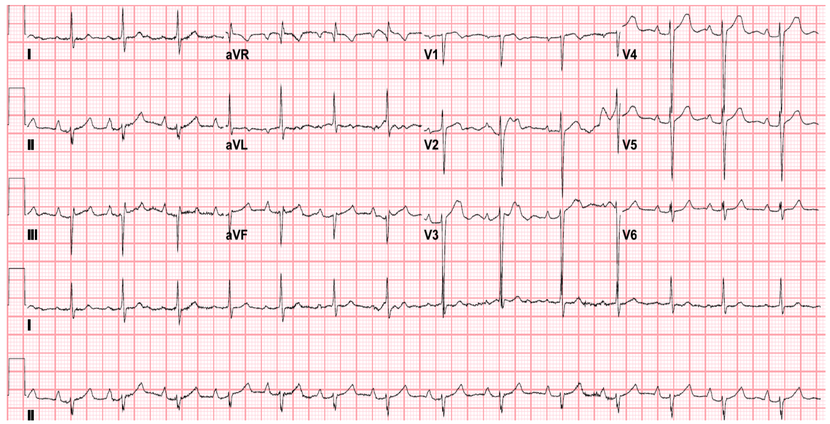

Figure 1 Baseline ECG one year prior. September 2019

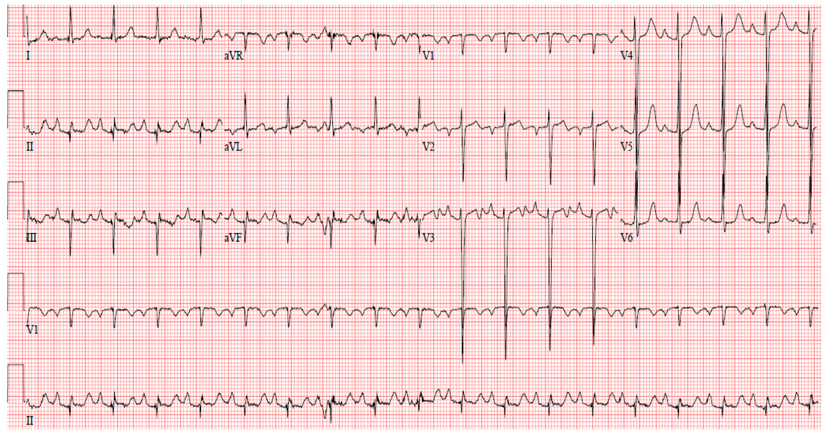

Figure 2. ECG in ER after unresponsive episode. Sinus bradycardia. First degree AV block. May 22 2020

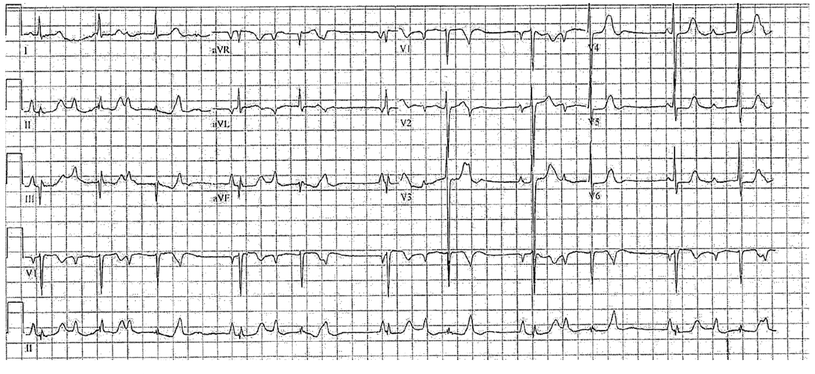

Figure 3. ECG Second Degree AV block (Wenckebach.) May 26 2020

ECG Progression

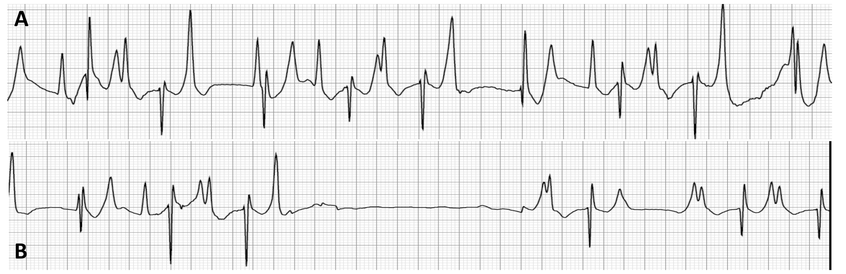

Figure 4 A/B. Holter Monitor. (A)Second degree AV block, Mobitz type 1, blocked sinus beats with deceleration of sinus rate, suspicious of infrahisian heart block and (B)Rare third degree heart block January 2021

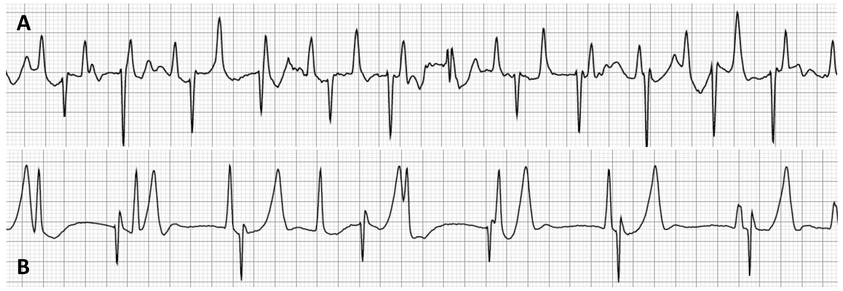

Figure A/B 5. Holter monitor after sedation March 2021. (A) High grade heart block with intermittent AV conduction (B) Complete heart block

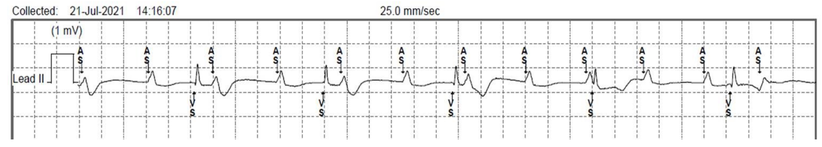

Figure 6 Pacemaker Check demonstrating persistent 3rd degree heart block (atrial sinus rate of 105 bpm with narrow complex ventricular escape of 48 bpm

Discussion and Conclusion

Our patient experienced progressive infrahisian conduction deficit over 7 months eventually requiring a pacemaker. It is our suspicion that his initial episode of unresponsiveness may have been due to yet undiagnosed conduction disease rather than beta blockade ingestion. He had no overt symptoms per the family, although has been noted to have increased energy since the procedure. Diligence in monitoring the electrophysiologic health of these rare patients is essential.

Acknowledgments

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