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Visual Diagnosis in Emergency Medicine

SPONTANEOUS RETURN OF SINUS RHYTHM IN A CHILD BORN WITH COMPLETE HEART BLOCK

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CASE REPORT

A 7-year-old girl presented with late recovery of sinus rhythm from an isolated autoantibody-related congenital complete atrioventricular block (AVB). She was born to a healthy, primigravida, asymptomatic 29-year-old woman with undiagnosed Sjögren syndrome (positive anti–Sjögren syndrome-related antigen A [anti-SSA] and SSB). Because of complete AVB with persistent profound bradycardia that was unresponsive to treatment with intravenous immunoglobulin G (IVIG) and because of the development of low cardiac output, an epicardial, single-chamber, rate-responsive, permanent pacemaker (SESR01; Medtronic Inc., Minneapolis, MN) was implanted when the child was 1 week old, and the pacemaker was programmed to VVIR mode with a heart rate limit of 100–150 beats/min. The pacemaker was placed in an abdominal pocket and the epicardial pacing lead was positioned on the right ventricular free wall.

Lead fracture with complete separation (Figure 1B) occurred while bending over backward during play at about 5 years of age. The electrocardiogram (ECG) showed a normal sinus rhythm at that time. Because she was asymptomatic and because she had a normal sinus rhythm, we left the pacemaker alone and did not change the initial pacemaker's setting (pacing rate limit of 100–150 beats/min).

At 7 years of age, the patient presented to our facility when she suddenly felt slow and regular muscle twitching in her abdominal wall, which was initiated by the fractured pacer wire. Her ECG showed pacing spikes created by the pacemaker output at 65 ppm, which were not followed by a QRS, and which were asynchronous with the normal intrinsic beats of 86 beats/min (Figure 1C). The pacemaker's generator and wire were removed surgically after this event.

DISCUSSION

This unusual presentation demonstrates an ECG with pacing spikes created by the pacemaker output at 65 ppm, which were not followed by a QRS, and were asynchronous with the normal intrinsic beats of 86 beats/min. Lead fracture with the failing battery having reached the elective replacement indicator (ERI) may explain why the pacemaker delivered electrical stimuli at a fixed rate and failed to capture despite the patient's intrinsic cardiac activity. The ERI of a pacemaker is intended to be used as a strict criterion for the appropriate timing of pulse generator replacement.

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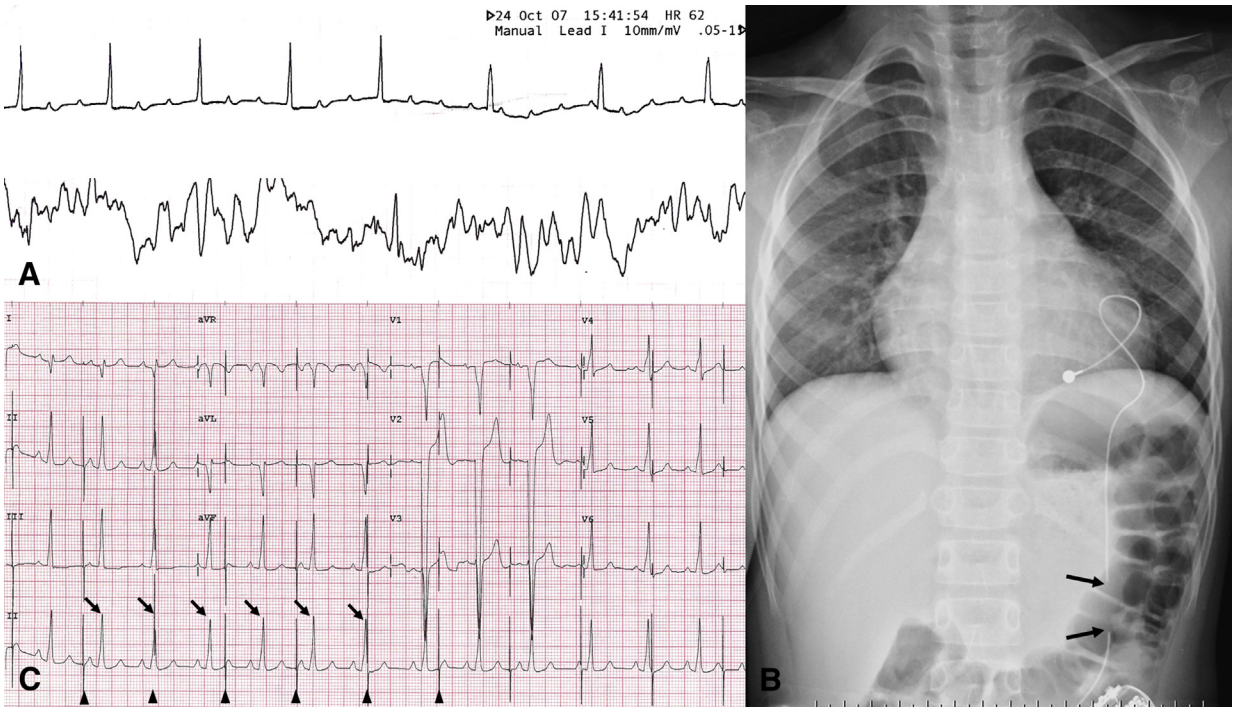

Figure 1. (A) Electrocardiogram showing a complete atrioventricular block with a ventricular rate of 62 beats/min (top) and ventricular fibrillation (bottom). (B) Chest roentgenography revealed pacing lead fracture with total disconnection (arrows). (C) The spikes (arrowheads) created by the pacemaker's output are not followed by a QRS and are asynchronous with the normal intrinsic beats (arrows) on the electrocardiogram.

Autoantibody-related congenital compete AVB is a rare neonatal disease with an overall prevalence of 1 in 20,000 live births. AVB affects approximately 2% of fetuses born to mothers with anti-SSA or anti-SSB antibodies (1). Most affected newborns have a potentially lethal complete AVB, which is irreversible, and approximately 57% to 66% of children born alive with complete AVB require a permanent pacemaker before reaching adulthood (2–4). Although a pacemaker can prevent electrophysiological heart failure, it cannot prevent myocardial injury induced by persistent maternal autoantibodies that may manifest clinically as irreversible AVB and myocardial dysfunction after birth (2–5). IVIG can facilitate anti-idiotype regulation and modulate the response of macrophages with secondary reduction of inflammation and fibrosis of the heart. Because this patient received IVIG in the neonatal period, a possible reason that postnatal IVIG treatment was beneficial in terms of heart rate improvement and absence of dilated cardiomyopathy was that it accelerated the clearance of potentially pathogenic autoantibodies, thereby mitigating the risk of myocardial injury progression. Close long-term follow-up is warranted in such cases and further studies are needed to elucidate the precise underlying mechanisms involved.

REFERENCES

1. Buyon JP, Clancy RM. Neonatal lupus: basic research and clinical perspectives. Rheum Dis Clin North Am 2005;31:299–313.
2. Di Mauro A, Caroli Casavola V, Favia Guarnieri G, et al. Antenatal and postnatal combined therapy for autoantibody-related congenital atrioventricular block. BMC Pregnancy Childbirth 2013;13:220.
3. Strasburger JF, Wakai RT. Fetal cardiac arrhythmia detection and in utero therapy. Nat Rev Cardiol 2010;7:277–90.
4. Hon KL, Leung AK. Neonatal lupus erythematosus. Autoimmune Dis 2012;2012:301274.
5. Udink ten Cate FE, Breur JM, Cohen MI, et al. Dilated cardiomyopathy in isolated congenital complete atrioventricular block: early and long-term risk in children. J Am Coll Cardiol 2001;37:1129–34.