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Long QT syndrome with AV Wenckebaching & bundle branch block in a neonate

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ABSTRACT

We present a case of 21-day-old neonate brought with history of 3 episodes of syncope. Evaluation revealed congenital long QT syndrome associated with long cycle atypical AV Wenkebaching with a long short cycle sequence related left bundle branch aberrancy. Syncope was attributed to multiple episodes of Torsades de Pointes, necessitating emergency epicardial pacemaker implantation. In addition, child was started on oral propranolol therapy. On 2 months follow up, child was stable with no ventricular high rate episodes during pacemaker interrogation.

1. Introduction

Congenital long QT syndrome (LQTS) is occasionally complicated by impaired atrio-ventricular (AV) conduction, mostly in the form of 2:1 AV block (pseudo AV block). This form of LQTS can manifest before birth or during neonatal life and has been associated with a guarded prognosis [1].

We report a case of congenital LQTS with variable AV conduction abnormalities which was successfully treated with epicardial pacemaker implantation.

2. Case report

A 21-day-old neonate was brought to emergency with history of 3 episodes of sudden onset unresponsiveness with cyanosis in last 10 days. The episodes were not associated with excessive crying and resolved spontaneously within a minute of onset. Physical examination, complete blood count, sepsis screen and basic metabolic profile were unremarkable. 12-lead ECG analysis revealed sinus rhythm with predominant Mobitz Type I second degree AV block with alternating 3:2 and 2:1 conduction, with intermittent advanced AV block, along with a prolonged QTc of 592ms (Fig. 1). Although, genetic analysis could not be performed, a diagnosis of type 3 LQTS was suggested by the ECG pattern of late-onset peaked T waves. There was intermittent long cycle (8:7) atypical AV Wenckebach phenomenon as well (Fig. 2), (Supplementary Fig. 1). The first beat of each cycle appeared to have a normal QRS morphology but the second beat onwards, the QRS morphology changed to an incomplete left bundle branch block (LBBB) pattern. This was triggered by a long pause due to non-conducted P wave which lead to a long-short cycle sequence leading to cycle length dependent prolongation of refractory period of left bundle (Ashman phenomenon). The subsequent incomplete LBBB persisted for six beats as in Fig. 2 till the last P wave was blocked in the Wenckebach sequence. This likely occurred because of retrograde transseptal activation of the left bundle from the right bundle. 2-D transthoracic echocardiography revealed a structurally normal heart. There was no history of any antenatal drug intake by mother, and her past medical history was insignificant. There was no family history of sudden unexplained death, syncope, drowning or deafness. ECGs of both parents were normal, with normal QT intervals. On further investigations, mother was found to be positive for anti-Ro antibodies (Observed value – 36.8RU/ml, Normal <15RU/ml).

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Abbreviations

AV	Atrio-ventricular
ECG	Electrocardiograph
LBBB	Left Bundle branch block
LQTS	Long QT syndrome
Ms	milliseconds
mV	millivolt
TdP	Torsades De Pointes
VT	Ventricular Tachycardia

On the day of admission, baby had multiple episodes of syncope due to non-sustained Torsades de pointes (Fig. 3a) with no response to magnesium sulfate and hence, underwent epicardial pacemaker implantation on an emergent basis. Implantable cardioverter-defibrillator (ICD) was not implanted due to the babys' age and weight. The two poles of a Medtronic epicardial bipolar lead (4968) were sutured to the right ventricular outflow tract and anterior surface of the right ventricle, respectively, with placement of an Effecta SR (Biotronik Inc., Berlin, Germany) generator behind the rectus muscle. Implant parameters were as follows – pacing threshold 1.2V @ 0.5ms, R wave 19mV and pacing impedance 632Ω. The pacing output was set at 4V @ 0.5ms and the lower/upper rates were set at 120/160 bpm. Following pacemaker insertion, oral propranolol at 2mg/kg/day was started. No block, TdP or syncope occurred in the following 7 days of hospital stay (Fig. 3b), hence the baby was discharged. On follow up visit at 2 months, child was stable and no ventricular high rate episodes were observed during pacemaker interrogation.

3. Discussion

LQTS in association with 2:1 AV block is an unusual type of LQTS, accounting for approximately 5% of the total cases and has a high mortality rate [2]. Mutation in HERG (LQTS2), SCN5A (LQTS3), CACNA1 (LQTS8), and SCN4B (LQTS10) have been associated with this abnormality [3]. LQTS with AV Wenckebaching has been rarely

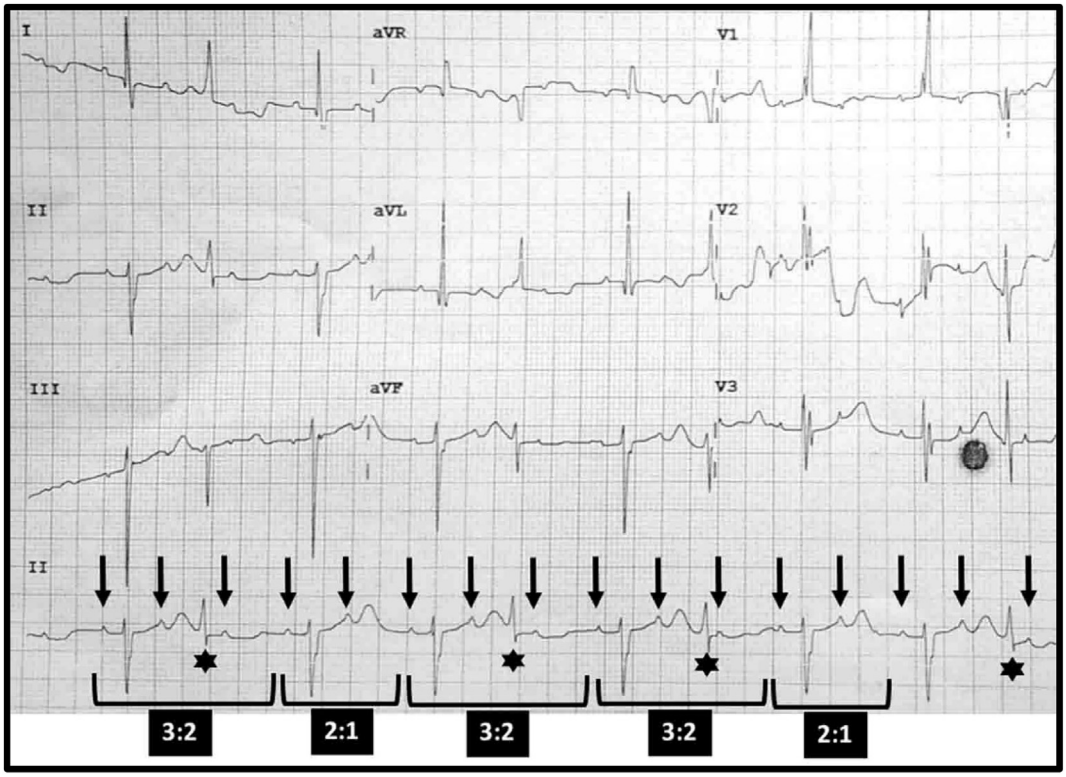

Fig. 1. ECG showing equally spaced P waves (Black arrow) with Mobitz Type I second degree AV block with alternating 3:2 and 2:1 conduction along with a prolonged QTc of 592ms. Also note intermittent QRS with incomplete left bundle branch block (Black star).

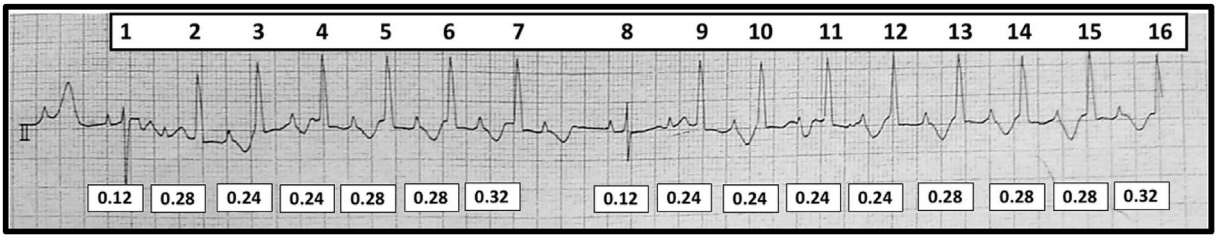

Fig. 2. Rhythm strip showing long cycle AV Wenckebach phenomenon with first QRS complex of each cycle (1,8) having 'normal' morphology with rest of the following complexes showing incomplete left bundle branch block caused by Ashman phenomenon.

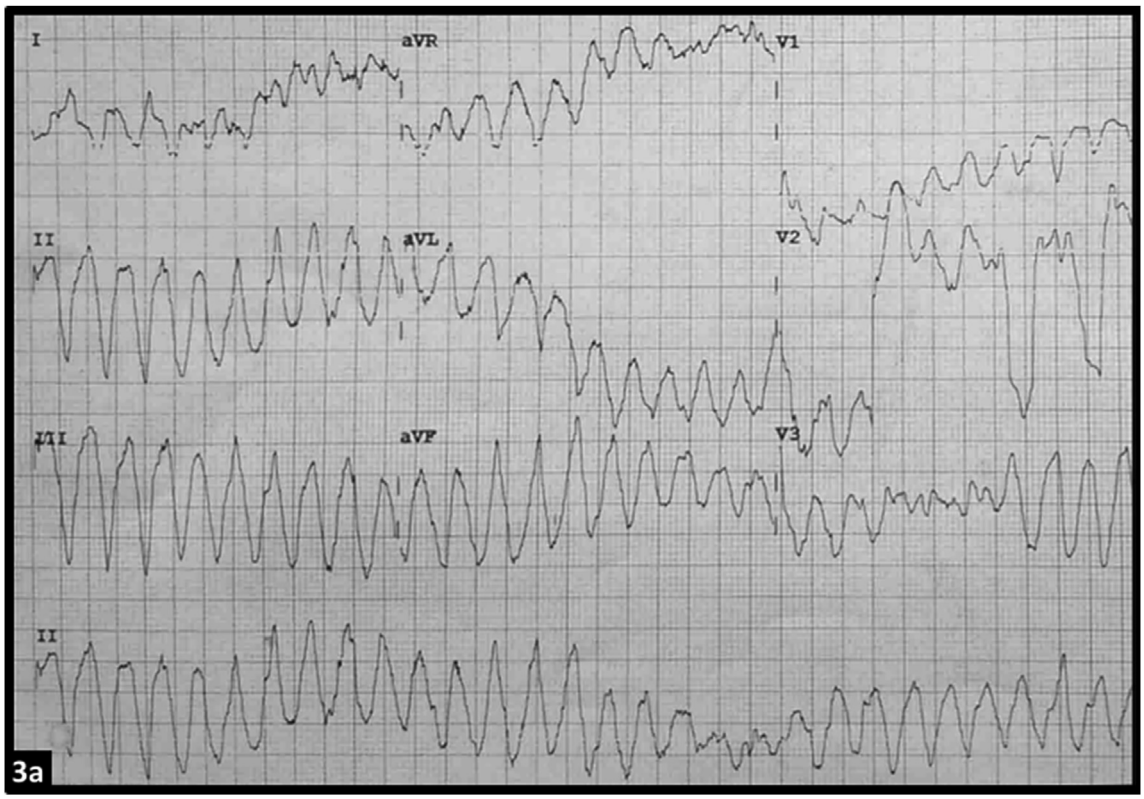

Fig. 3a. ECG showing polymorphic ventricular tachycardia (Torsades de pointes).

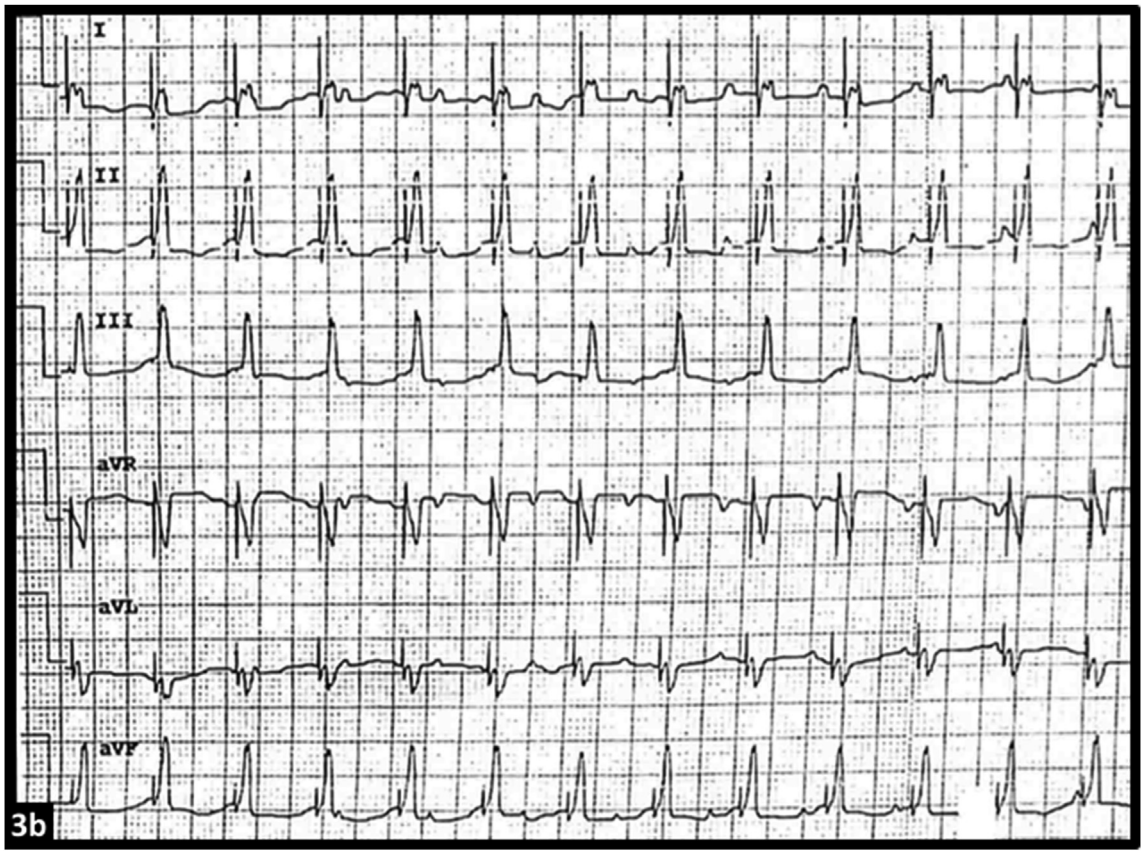

Fig. 3b. ECG after pacemaker implantation showing paced rhythm.

reported.

In the current case, the site and mechanism of conduction block could not be confirmed due to hemodynamic instability precluding an invasive electrophysiological testing. Nevertheless, AV node is the likely site of block as AV Wenckebaching is a far more common manifestation of AV nodal block than isolated His Purkinje system involvement. AV Wenckebaching due to His-purkinje system involvement is associated with subtle changes in PR interval and/or unaccounted changes in QRS axis [4]. But as seen in Fig. 2, changes in PR interval are not subtle (Range – 0.12 to 0.32s), and the change in QRS axis can be explained by functional aberration (incomplete LBBB) secondary to Ashman phenomenon [5]. Presence of Wenckebach phenomenon might reflect a relatively milder degree of AV nodal conduction disturbance. The markedly prolonged QTc and the frequent episodes of Torsades de Pointes favour LQTS with 'pseudo' AV block as the primary mechanism of AV block. An alternative explanation could be 'true' AV nodal block with associated long QTc, due to injury to AV node and myocardial ion channels by anti-Ro antibodies [6]. In the current case, the maternal Anti-Ro antibody titer was at best mildly elevated (36.8 RU/ml), although, was measured in the postnatal period. Hence, evidence is insufficient in the present case to implicate Anti-Ro positivity as a cause of AV nodal block.

In patients with markedly prolonged QT interval at a slow heart rate, beta-blockers may further prolong QT interval by slowing the heart rate [8]. Hence, we were sceptical to start beta blocker in our patient who was bradycardic due to intermittent AV block. Mexiletine has also been used to treat patients with LQTS and impaired AV conduction, however the response has been heterogenous [7,8].

Device implantation (pacemaker or ICD) should be considered in patients having persistent TdP episodes despite medical therapy. Permanent pacing is reasonable in patients with LQTS and advanced AV block, especially in LQTS3 [9]. The beneficial effects have been attributed to shortening of QTc by promotion of more homogeneous repolarization, thus reducing the frequency of recurrent syncopal events. The relative efficacy of atrial, ventricular, or dual-chamber pacing in preventing recurrent TdP in these patients is not well established. In situations where permanent pacing is not available, temporary pacing can be useful to tide over the crisis period. ICD implantation is recommended for selected patients of LQTS who are survivors of sudden cardiac arrest or who have sustained ventricular arrhythmias/recurrent syncope despite drug therapy [10]. Infants are particularly at risk for injury to the ICD either through somatic growth or repetitive movement. Further, the risk of inappropriate ICD shocks due to lead malfunction, atrial arrhythmias or sinus tachycardia is reportedly high in infants. Therefore, the final decision to implant a permanent pacemaker or an ICD rests at the discretion of treating physician, keeping in mind the age and weight of the child, and the respective institutional experience.

4. Conclusion

Congenital LQTS should be suspected when intermittent AV block alternating with VT is present. Implantation of a pacing system should be considered in LQTS associated with AV block if TdP episodes occur despite medical therapy.

Credit author statement

Jay Relan: Clinical management and follow up, original draft preparation.

Jaskaran Singh Gujral: Draft review, clinical management and follow up.

Seemala Saikrishna Reddy: Draft review, clinical management.

Sukhjeet Singh: Draft review, clinical management.

Neeraj Parakh: Supervision, Writing- Reviewing and Editing, Validation.

Sivasubramanian Ramakrishnan: Clinical management, Writing- Reviewing and Editing.

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Declaration of competing interest

'The Author(s) declare(s) that there is no conflict of interest'

"All authors have read and approved the manuscript."

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ipej.2020.07.005.

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