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

Jay Relan, Jaskaran Singh Gujral, Seemala Saikrishna Reddy, Neeraj Parakh*, Sukhjeet Singh, Sivasubramanian Ramakrishnan
All India Institute of Medical Sciences (AIIMS), New Delhi, India

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).
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 baby’s 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

---

**Abbreviations**

| Abbreviation | Description                  |
|--------------|------------------------------|
| 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      |

---

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.
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 Wenckebach, 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 heterogeneous [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 syncpe 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.
Sivasubramanian Ramakrishnan: Clinical management, Writing- Reviewing and Editing.

Funding support

None.

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.

References

[1] Aziz PF, Tanel RE, Zelister IJ, Pass RH, Woand TS, Vetter VL, et al. Congenital long QT syndrome and 2:1 atrioventricular block: an optimistic outcome in the current era. Heart Rhythm 2010 June;7:781–5.
[2] Anuwutnavin S, Wanitpongpan P, Chungsomprasong P, Soongswang J, Srisantitoj R, Wataganara T. Fetal long QT syndrome manifested as atrioventricular block and ventricular tachycardia: a case report and a review of the literature. Pediatr Cardiol 2013 Dec;34:1955–62.
[3] Medeiros-Domingo A, Iturrade-Torres P, Ackerman MJ. Clinical and genetic characteristics of long QT syndrome. Rev Esp Cardiol 2007 Jan;60:739–52.
[4] Akhtar M. Wenckebach phenomenon in the His-purkinje system. Card Electrophysiol Clin 2016 Dec;8(4):767–8.
[5] Lukić I, Mahović D, Silvjak V. Ashman phenomenon: an often unrecognized entity in daily clinical practice. Acta Clin Croat 2010 Mar;49(1):99–100.
[6] Cimaz R, Stramba-Badiale M, Brucato A, Catelli L, Panzeri P, Meroni P. QT interval prolongation in asymptomatic anti-SSA/RS-positive infants without congenital heart block. Arthritis Rheum 2000 May;43(5):1049–53.
[7] Sarubbi B, Frisso G, Romeo E, Evangelista E, Cordella A, D’Alto M, Santarpia G, Russo MG, Salvatore F, Calabro R. Efficacy of pharmacological treatment and genetic characterization in early diagnosed patients affected by long QT syndrome with impaired AV conduction. Int J Cardiol 2011 May;149(1):109–13.
[8] Li G, Zhang L. The role of mexiletine in the management of long QT syndrome. J Electrocardiol 2018 Nov;51(6):1061–5.
[9] Viskin S. Cardiac pacing in the long QT syndrome: review of available data and practical recommendations. J Cardiovasc Electrophysiol 2000 Mar;11(5):593–9.
[10] Epstein AE, DeMarco JP, Ellenbogen KA, Estes NANM III, Friedman RA, Gettes LS, et al. 2012 ACCF/AHA/HRS focused update incorporated into the ACCF/AHA/HRSC 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol 2013;61(3):c6–75.