Cardiology in the Young

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Favourable outcome of a severe bradyarrhythmia in a neonate: a case report

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Abstract

We report the case of a term neonate with severe fetal bradycardia with an unusually benign clinical course with follow-up till infancy.

Brief Report

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Complete congenital heart block (CCHB) is an irreversible immune-mediated bradyarrhythmia usually presents with a structurally normal heart (isolated complete congenital heart block) occurring in 1 in 20,000 live births and can be life-threatening in the presence of severe bradycardia, dilated cardiomyopathy, or endocardial fibroelastosis.[1] The overall mortality is 20–30% requiring pacing in 60% during infancy.[2] Herein, we describe the clinical course of a life-threatening arrhythmia with severe bradycardia with a favourable outcome.

Clinical description

An infant weighing 3204 g was born to a 29-year-old mother at 38^{+5} weeks of gestation. She was registered at a local hospital with irregular follow-up and had no history of diabetes or hypothyroidism. Antenatal ultrasound at 21 weeks showed no abnormalities with a fetal heart rate of 152 beats per minute (bpm). However, ultrasonography at 37 weeks detected mild cardiomegaly and fetal bradycardia with a ventricular rate of 94 bpm with ectopic beats, and an atrial rate of 134 bpm, with possible atrio-ventricular conduction abnormality. The mother was referred to our centre and a fetal echocardiography (38^{+4} weeks) confirmed bradycardia (43 bpm), though without atrio-ventricular discordance, and was planned for further evaluation. On enquiry, there was no history of fever, joint pain, rash, ocular dryness, and oral ulcers, or family history of autoimmune disease. The following day, she delivered a male infant vaginally who cried at birth, with apgar scores of 8 and 8 at 1 and 5 minutes, respectively. Delivery room evaluation confirmed neonatal bradycardia (50–55 bpm) with normoxemia.

In the NICU, the neonate was euthermic, pink, without respiratory distress, and with heart rate of 50–60 bpm without any variability to stimulation. His saturations were 96–98% and blood pressure was normal. All peripheral pulses were well felt and capillary refill time was <3 seconds. The infant had normal heart sounds, with no murmur, hepatomegaly, or signs of hydrops. Chest radiography showed a normally sized and shaped heart. An electrocardiogram showed complete congenital heart block with an atrial rate of 110 bpm and a ventricular rate of 50 bpm with narrow complex escape (Fig 1). Echocardiography showed two small ostium secondum atrial septal defects and a 2-mm patent ductus arteriosus with left to right shunt with mild tricuspid regurgitation and good biventricular function without endocardial fibroelastosis or dilated cardiomyopathy. A cardiologist and electrophysiologist opined and advised close follow-up with emergency pacing in case of hemodynamic failure.

Subsequently, maternal evaluation showed microcytic hypochromic anaemia with iron deficiency, raised erythrocyte sedimentation rate and C-reactive protein, with absent urinary proteins/blood. Lupus anticoagulant, anti-phospholipid, cardiolipin, and beta2-glycoprotein antibodies were negative. She tested positive for Sjogren's syndrome-related antigen A (SS-A) native (60 kDa), SS-A/Ro-52 recombinant and SS-B antigen, and negative for dsDNA, ribonucleoprotein/Sm, Scl-70, Jo-1, nucleosomes, histones, centromere B, and ribosomal-p protein by anti-nuclear antibody Blot method. Maternal electrocardiogram and Schirmer test were normal. Direct coombs test was negative and TSH level was 3.2 mIU/ml.

The infant was stable throughout hospitalisation and was successfully discharged on day 12. At 14 months follow-up, the infant is healthy with heart rates of 60–65 bpm, without signs of cardiac failure, need of pacing, or hospitalisation. The repeat echocardiography revealed good biventricular function and Holter ECG monitoring suggested complete AV block with an average heart rate of approximately 76/min with minimal heart rate of 39/min with QTc average of 455 ms, without any evidence of wide QRS complexes of escape rhythm, ventricular arrhythmia, or long pause (Fig 2). Both mother and infant remain under close surveillance.

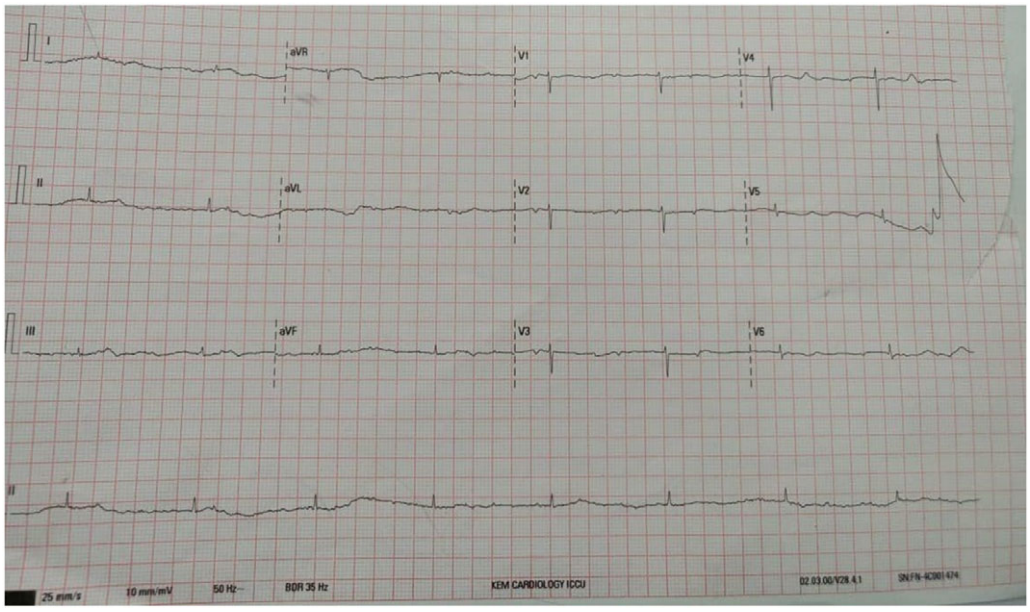

Figure 1. 12-lead electrocardiogram tracing of the neonate on day 1 of life.

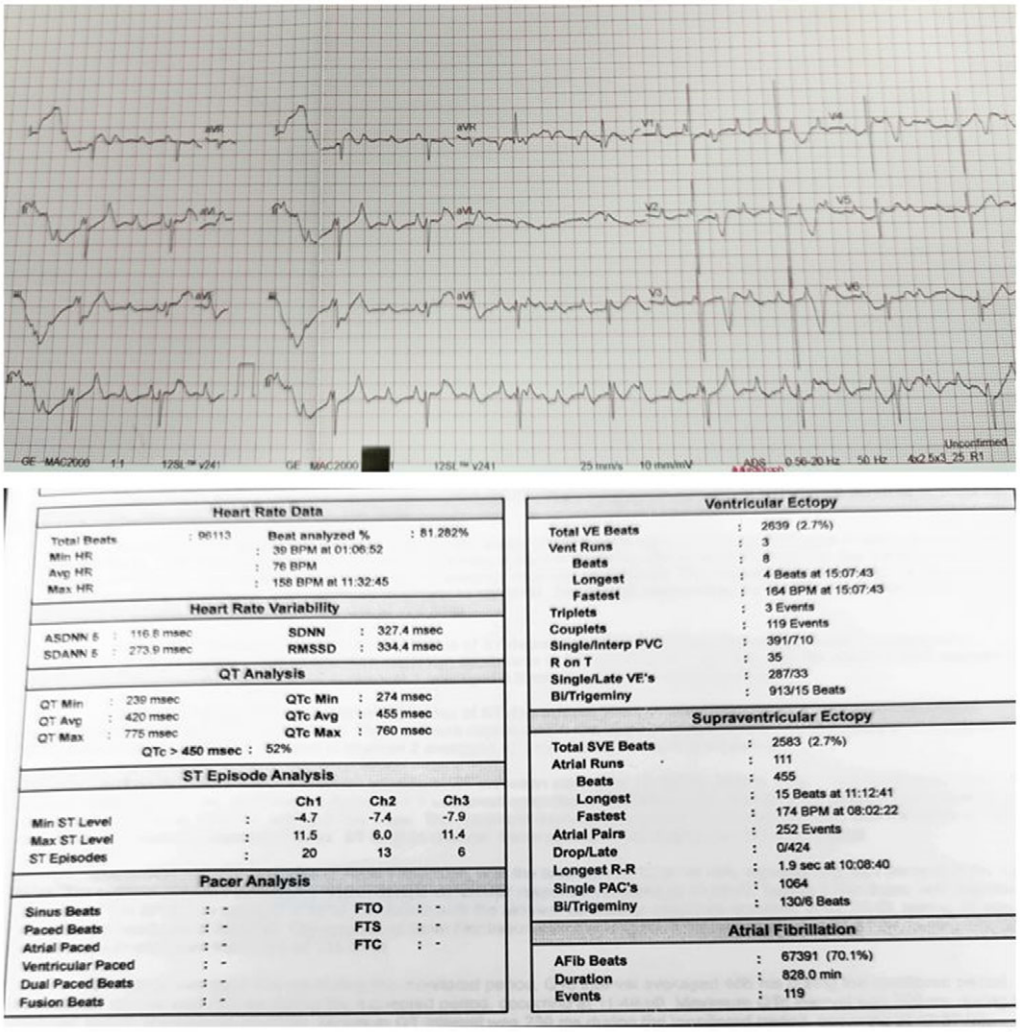

Figure 2. 12-lead electrocardiogram tracing (above) and Holter electrocardiogram (below) on follow-up at 14 months of age.

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Discussion

Complete congenital heart block is the most common and severe atrio-ventricular block usually detected in utero, at birth or within the first month.[3] Risk factors include maternal diseases (diabetes mellitus), viral infections (coxsackie, adenovirus), and drugs (anti-epileptics, lithium). Fetal factors such as cardiac malformations and genetic channelopathies are also implicated. Isolated CCHB is more common in infants of mothers with autoimmune disease and/or auto-antibodies positivity. Asymptomatic antibody carrier status in mother of index case was similar to a systematic review of 856 mothers, where over 50% were asymptomatic despite having anti-Ro and anti-La antibodies.[3] These mothers could be immunologically positive even years before clinical symptoms appear, making a definitive diagnosis of SLE or Sjogren difficult. However, risk of recurrence is nearly 20% in subsequent pregnancies especially when both auto-antibodies are present,[2] necessitates strict follow-up. Auto-antibodies transferred trans-placentally impede clearance of apoptotic fetal cardiocytes, causing inflammation, fibrosis, and rhythm impairment.[4] Often non-cardiac manifestations involving blood, liver, and skin are observed, which resolve at approximately 6 months postnatally, coinciding with waning maternal autoantibodies from infant's circulation.[2] Our infant showed none of these manifestations. To decrease the recurrence risk of CCHB in mothers with autoantibodies, use of hydroxychloroquine and fluorinated steroids in antenatal period has been demonstrated, however, current recommendations emphasise on serial echocardiograms every 1-2 weeks from 16th week gestation, to detect premature atrial contractions and moderate pericardial effusion and then initiate preventive therapies.[5] IVIG used in a recent trial also showed no reduction in recurrence.[6]

Most cases are diagnosed in the fetal period, with 82% before 30 weeks of gestation.[2] Antenatal therapeutic strategies include maternal steroids, sympathomimetics, and plasmapheresis.[7] In our case however, due to irregular antenatal visits, the detection was possibly delayed with no window for antenatal intervention. Shokrzadeh performed longitudinal analysis of fetal heart rates and showed an increased risk of perinatal death, severe fetal bradyarrhythmia, and need of emergency pacing with ventricular rates below age-specific mean.[8] A ventricular rate <55 bpm is considered critical with guarded fetal prognosis.[3] Our neonate, despite having a nadir of 43 bpm, did not develop hydrops, and had an uncomplicated perinatal course, similar to a few case reports in literature.[9] Tunaoglu et al described a term neonate where the nadir heart rates were 39 bpm and pacing was not required till reporting at 8 years.[10]

The overall mortality of CCHB is approximately 20–30%, with upto 70% deaths in utero.[2,3] Risk factors for mortality include fetal diagnosis, hydrops, endocardial fibroelastosis, and delivery ≤32 weeks.[11] The mortality in the presence of either dilated cardiomyopathy or endocardial fibroelastosis is >50%, increasing to 100% if both are present. Pacemaker insertion is recommended in asymptomatic cases with ventricular rate <50 bpm, broad QRS escape rhythm, prolonged QT interval, or complex ventricular ectopy. Haemodynamic instability with ventricular dysfunction attributable to bradycardia mandates emergency pacing.[12] Buyon et al reported 63% of 107 children born alive had pacemakers inserted, of which 35 required it within the first 9 days of life, while 15 additional were paced in infancy, and 17 after 1 year. This highlights the ability of some children to tolerate bradycardia quite well for some time, while others require cardiac pacing in the first few days of life and decision of permanent pacing requires detailed diagnostics.

Our infant is currently 14 months old and doing well without pacing.

Conclusion

Complete congenital heart block with severe bradycardia (<55 bpm) can have a benign clinical course, though strict vigilance is mandatory. Detection of auto-antibodies and follow-up of these mothers is imperative as it has implications in future pregnancies.

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Conflict of interest. None.

Ethical standards. None.

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