

# Remission of congenital complete heart block without anti-Ro/La antibodies: A case report

Souvik Mitra, Anindya Kumar Saha, Syamal Kumar Sardar, Arun Kumarendu Singh

Department of Neonatology, Institute of Postgraduate Medical Education and Research, SSKM Hospital, Kolkata, West Bengal, India

## ABSTRACT

Anti-Ro/La negative congenital heart block (CHB) is uncommon. We report one such case of CHB, with no associated structural heart disease or maternal autoantibodies. The heart block reverted to sinus rhythm spontaneously at two weeks of age, and the patient remains in sinus rhythm at a one year followup. Whether patients with antibody negative complete heart block have a different clinical course is conjectural.

**Keywords:** Anti Ro/La antibody, congenital heart block, neonatal bradycardia

## INTRODUCTION

Congenital atrioventricular (AV) block is a rare but important cause of neonatal bradycardia, which occurs in approximately 1: 15,000-1: 22,000 live births.<sup>[1]</sup> Isolated congenital heart block (CHB) is mainly associated with the neonatal lupus syndrome, but it can be observed in children whose mothers have other connective tissue diseases, such as Sjögren's syndrome. Nearly half of the mothers do not have connective tissue disease when their children are born with CHB, but most of them have anti-Ro/La autoantibodies.<sup>[2]</sup> Anti Ro/La negative cases constitute around 30% of all cases of CHB, which are mostly associated with structural heart defects.<sup>[3]</sup> There have been few reports of non-immune CHB without associated structural heart defects, with their prognoses being largely variable. We report a case of congenital AV block, with no associated structural heart disease or maternal autoantibodies.

## CASE REPORT

A single-term, appropriate-for-date, girl baby, with birth weight 2478 g, born out of a non-consanguineous marriage to a 24-year-old primigravida mother, presented at birth with a heart rate of 45/minute. The

baby was born by normal vaginal delivery and had APGAR scores of 7 and 8 at one and five minutes. The obstetric history revealed documented fetal bradycardia during labor, but normal heart rate on the ultrasound scans at 18 and 35 weeks of gestation. The mother had no history of gestational diabetes, hypertension, hypothyroidism, any fever with rash, or drug intake during pregnancy, nor did she have any symptoms suggestive of connective tissue disease. There was no history of cardiac disease in the family.

At birth, the baby was otherwise healthy with a heart rate of 45/minute, without any symptoms of congestive cardiac failure. Serum electrolytes were normal and echocardiography revealed no structural heart disease. The baby was put on continuous electrocardiographic monitoring. The electrocardiogram (EKG) on day one revealed complete AV dissociation with an atrial rate of 115/minute, ventricular rate of 45/minute, QRS duration of 0.08 seconds (occasionally > 0.08 seconds), corrected QT interval (QTc) of 0.48 seconds, and an left bundle branch block (LBBB) pattern [Figure 1]. EKG on day three revealed no AV dissociation with both atrial and ventricular rates of 93/minute and QRS duration of 0.08 seconds [Figure 2]. Although the EKG reverted to sinus rhythm, the PR interval (0.18 seconds) was prolonged, the QRS duration was at the upper limit of normal, and the EKG showed a right bundle branch block (RBBB) pattern. There was marked ST depression in V1 and marked right axis deviation, indicating normal right ventricle (RV) dominance in the newborn period. The EKG on day four showed a similar picture as on day three with a rate of 107/minute. On day six, however, the bradycardia again worsened. This time the ventricular rate varied between 45 and 60/minute, atrial rate was

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**Address for correspondence:** Dr. Souvik Mitra, 618, Block "O", New Alipore, Kolkata - 700 053, West Bengal, India. E-mail: souvik4sps@yahoo.com

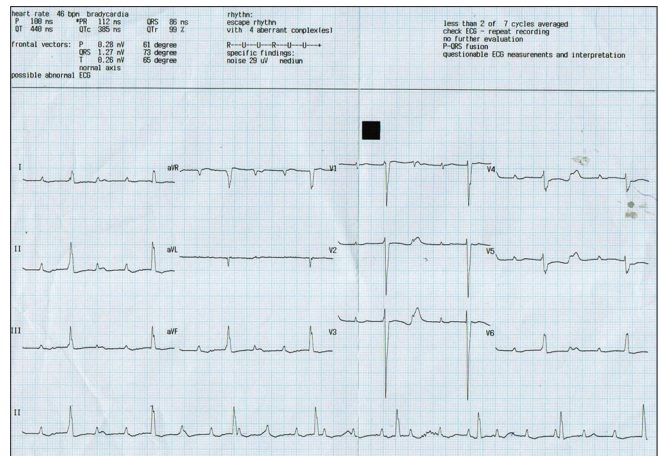
150/minute, and QRS duration was 0.08-0.1 seconds. This pattern prevailed over the next seven days. From day 13 onward, there was spontaneous improvement, with disappearance of AV dissociation and atrial-ventricular rates improving to 100/minute. The baby remained asymptomatic during the hospital stay. Cardiac function was regularly monitored using functional echocardiography, and the fractional shortening (FS%) was found to range between 32 and 38% during this period, which was normal for term neonates. Hence, cardiac pacing was withheld. The baby was discharged on day 29 of life. During this period, the heart rate varied between 85 and 120/minute, but no further episodes of AV dissociation were noted. The mother was tested for serum anti-nuclear antibody using the indirect immunofluorescence technique. Subsequently the mother's serum was subjected to antigen-specific enzyme immunoassay, using microtiter plates coated with a combination of antigens SSA/Ro, SSB/La, Sm, U1-RNP, Jo-1, and Scl70. The test results were negative.

The baby was followed up weekly for two months, then every fortnight for the next 14 months [Figure 3]. The baby did not develop any such episodes further. Serial echocardiography during this one-year follow-up revealed no evidence of any cardiac dysfunction. EKGs were done in both the parents and were found to be normal.

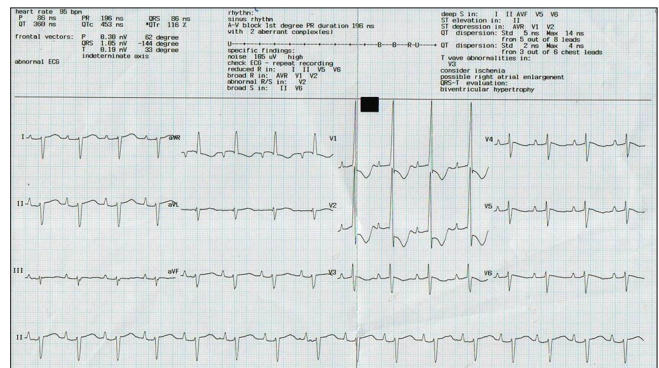
## DISCUSSION

Anti Ro/La negative CHB without structural heart defects is rare. The presence of maternal anti-Ro (SSA) or anti-La (SSB) antibodies have been demonstrated in most cases of isolated congenital AV blocks, with the presumed mechanism being that of immune complex deposition with specific damage to the fetal cardiac conduction system.<sup>[4]</sup> However, Brucato *et al.*, and Maeno *et al.*, observed CHB in 20 and 18% of mothers with anti-Ro/La negativity, respectively.<sup>[5,6]</sup> Among them, two fetuses had a second-degree AV block, one progressing to complete block soon after birth and the other at three months. The block alternated with normal sinus rhythm in the other two infants and reverted to a stable normal sinus rhythm in one patient.<sup>[5]</sup> Bruer *et al.*, reported a series of four patients where fetal heart block was found to occur in the absence of structural heart defects and maternal autoantibodies to SS-A/Ro and SS-B/La. These neonates were also found to exhibit an unstable pattern of AV block, with the blocks changing their degree and often reverting to sinus rhythm.<sup>[7]</sup> Our case exhibited a similar clinical course with an unstable pattern of AV block alternating with sinus rhythm.

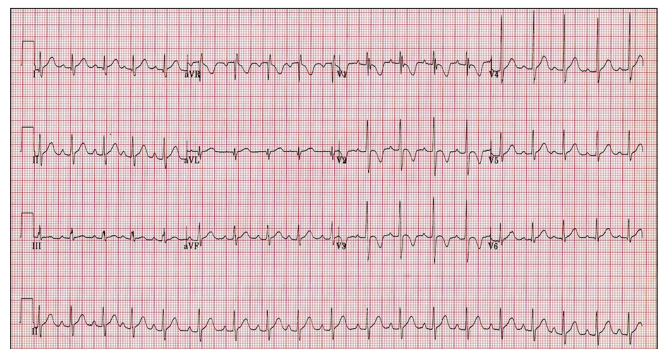
Interestingly, it has been found that anti-Ro/La negativity does not always rule out the possibility of an immune-mediated process. Although anti-SS-A (RO)



**Figure 1: Day one: Complete AV dissociation; atrial rate: 115/minute; ventricular rate: 45/minute; QRS duration: 0.08 seconds; QTc: 0.48 seconds; LBBB pattern**



**Figure 2: Day three: Sinus rhythm; no AV dissociation; PR interval: 0.18 seconds; QRS rate: 93/minute; QRS duration: 0.08 seconds; atrial rate: 93/minute; RBBB pattern with ST depression in V1 and right axis deviation**



**Figure 3: Fourteen months follow up: Normal sinus rhythm; Rate: 107/minute; Age-related normal findings of T-wave inversion in V1-V3 and RSR' pattern in V1**

antibodies/anti-SS-B (LA) usually maintain a stable profile for many years, late seroconversion has been observed, without a clear explanation.<sup>[8]</sup> Insensitive methods, a very low concentration of maternal antibodies, or even a true variation in antibody levels has been speculated to explain this phenomenon. Late seroconversion in those

patients reinforces the possibility of other unknown intrinsic (fetal) or extrinsic (maternal) factors involved in the genesis of the AV block, besides anti-SS-A (RO)/anti-SS-B (LA) antibodies. Moreover, other studies have proved that maternal antibodies are not sufficient to cause an AV block, because the prevalence is low in seropositive mothers, from 1 to 7.5%.<sup>[9]</sup>

There have also been conflicting reports regarding prognosis in such a group of patients. According to Berg *et al.*, the infant mortality rates were similar among mothers who had anti-Ro positivity, anti-Ro negativity, and children with CHB.<sup>[10]</sup> In a large multicentric study by Baruteau *et al.*, 79.4% required pacemaker implantation, but 90.1% of them had no systolic dysfunction on long-term follow-up.<sup>[11]</sup> Spontaneous reversal of a heart block and no cardiomyopathy on follow-up has also been documented by a number of reported series on such patients.<sup>[7,12,13]</sup>

In conclusion, the prognosis of congenital complete heart block in the absence of anti Ro/La antibodies may be relatively better. However, the entity may result from different causes. Further characterization of this subgroup is required.

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