DOI: 10.7860/JCDR/2025/78389.21901

Case Report

Paediatrics Section

Spontaneous Remission of Congenital Non-immune Mediated Heart Block in a Foetus at 21 Weeks Gestational Age: A Case Report

SREE DIVYA MOHAN[1], J JAYAKAVIYAH[2]

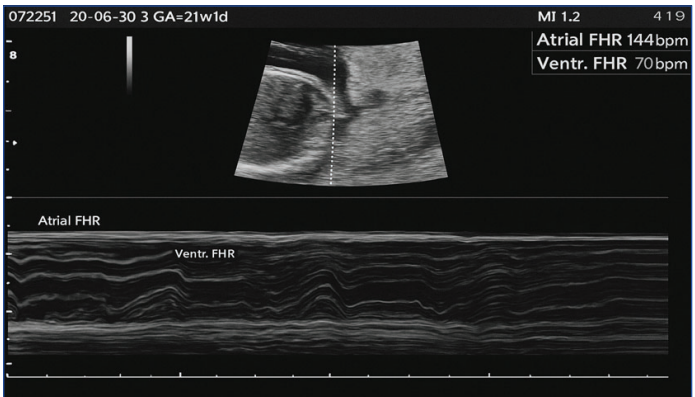
ABSTRACT

Isolated congenital Atrioventricular Block (AVB) in structurally normal hearts has an estimated prevalence of 1 in 15,000-22,000 live births. In non-immune cases, up to 50% are associated with structural abnormalities like atrial isomerism or congenitally corrected transposition of the great arteries. Rarely, AVB of unknown origin occurs in the absence of maternal antibodies, structural heart defects, trauma, metabolic disorders, or channelopathies. The natural history of idiopathic AVB remains poorly understood. A 29-year-old primigravida with no comorbidities was found to have foetal bradycardia, with atrial contractions at 144 beats per minute and ventricular contractions at 70 beats per minute during a routine anomaly scan at 21 weeks of gestation. Foetal Echocardiography (ECG) using M-mode confirmed a 2:1 AVB in an otherwise structurally normal heart. Maternal serology was negative for anti-SSA/Ro and anti-SSB/La antibodies. Repeat foetal ECG at 23 weeks showed spontaneous resolution to normal sinus rhythm. Serial prenatal assessments remained normal, and the pregnancy progressed without complications. At term (40 weeks), she delivered a healthy female neonate with a normal ECG and sinus rhythm. Postnatal follow-up confirmed normal cardiac function and development. This case underscores the potential for a benign outcome in isolated, non-progressive AVB in structurally normal hearts without maternal autoantibodies. Although the mechanisms of spontaneous resolution remain unclear, this case supports the importance of individualised evaluation and careful monitoring before initiating corticosteroids or invasive treatment. Additional case reports and prospective studies are vital to guide future management and counselling.

Keywords: Atrioventricular block, Echocardiography, Foetal bradycardia, Transient remission

CASE REPORT

A 29-year-old primigravida, non-consanguineously married, was referred at 21 weeks of gestation following the detection of foetal bradycardia during a routine anomaly scan. Foetal Echocardiography (ECG) using M-mode and pulsed-wave Doppler confirmed a 2:1 Atrioventricular Block (AVB), with atrial contractions at 144 beats per minute and ventricular contractions at 70 beats per minute [Table/Fig-1]. The foetal heart was structurally normal, with no evidence of hydrops foetalis. Echocardiographic assessments of both parents were also normal.



[Table/Fig-1]: M-mode foetal echocardiography showing atrial heart rate of 144 beats per minute and ventricular heart rate at 70 beats per minute at 21 weeks of gestation.

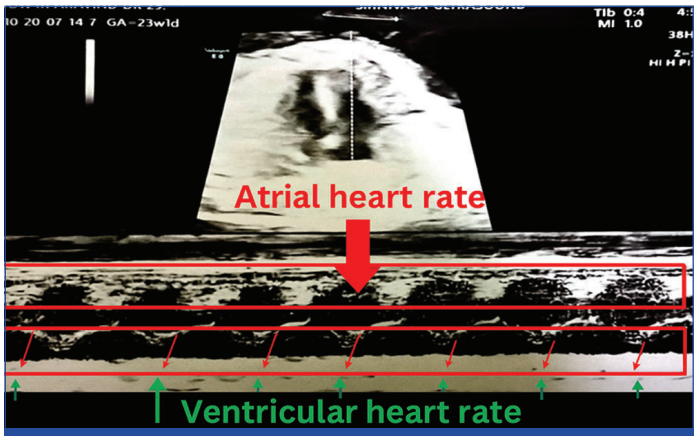
The maternal antenatal course was unremarkable. She was a non-smoker, non-alcoholic, had no comorbidities and was not on any medications other than routine iron and folic acid supplementation. There was no family history of sudden cardiac death, syncope, or bradyarrhythmias.

The differential diagnoses considered were Congenital Heart Block (CHB) associated with structural heart disease, such as left atrial isomerism and discordant AV connections, Immune-mediated AVB, particularly that resulting from maternal anti-Ro/SSA or anti-La/SSB antibodies, was also considered. In addition, genetic channelopathies were evaluated, including Long QT syndrome Type 3 (LQT3) and mutations in the SCN5A and NKX2.5 genes. A comprehensive autoimmune workup was performed. Maternal anti-SSA (Ro) and anti-SSB (La) antibodies were negative (4.49 U/mL and 2.62 U/mL, respectively, measured by enhanced ELISA). Additional serologic testing- including anti-Sm, RNP/Sm, Ro-52, Scl-70, PM-Scl 100, Jo-1, centromere proteins A and B, Proliferating Cell Nuclear Antigen (PCNA), dsDNA, nucleosome, histone, ribosomal P protein, and Anti-Mitochondrial Antibody (AMA)-M2 antibodies-was negative (immunoblot assay). ANA was also negative by immunofluorescence assay. These were effectively excluded based on a normal foetal ECG, negative maternal autoantibodies, and normal ECGs in both parents.

After consultation with a paediatric cardiologist and detailed counselling, the parents were informed about the potential progression of heart block, risk of hydrops foetalis, and the need for close monitoring. Given the absence of maternal autoantibodies, steroid therapy was not initiated. A repeat foetal echocardiogram was scheduled for follow-up.

Remarkably, at 23 weeks of gestation, repeat foetal ECG revealed restoration of normal AV conduction [Table/Fig-2]. Serial monthly foetal scans were performed thereafter and remained consistently normal throughout the pregnancy. At 40 weeks of gestation, the mother delivered a healthy 2,980 gm female neonate via spontaneous vaginal delivery. Postnatal evaluation showed a sinus rhythm at 140 bpm, with normal ECG and echocardiographic findings. The

neonate had no complications during the neonatal period. At two years of age, the child continues to demonstrate a normal sinus rhythm, with appropriate growth and developmental milestones.



[Table/Fig-2]: M mode foetal echocardiography showing foetal heart rate of 140 bpm at 23 weeks of gestation.

DISCUSSION

The occurrence of isolated CHB in foetuses with structurally normal hearts and negative maternal anti-Ro/La antibodies is rare, with an estimated prevalence of approximately one in 15,000 to 22,000 live births [1]. Despite its rarity, accumulating evidence from multiple studies suggests that non-immune-mediated CHB does occur and, in some instances, may resolve spontaneously.

For instance, Ju YT et al., reported a case of spontaneous resolution of complete AVB in a neonate with no detectable maternal autoantibodies or structural cardiac defects. The infant regained normal sinus rhythm five days after birth, thereby avoiding pacemaker implantation [2].

Shao S et al., found that 60% of foetuses with isolated, non-immune-mediated second-degree AVB experienced spontaneous reversion to normal sinus rhythm. Earlier gestational age at diagnosis and a higher atrial rate were significantly associated with favourable outcomes. Conversely, persistent AVB was suggestive of an underlying genetic condition, highlighting the importance of prenatal genetic testing to exclude heritable disorders such as long QT syndrome. These findings offer important guidance for clinical decision-making and prenatal counselling, allowing for tailored management strategies based on the potential for spontaneous recovery [3].

Supporting this, Maeno Y et al., reported that nine neonates (18%) in their multicentre cohort had CHB in the absence of maternal autoantibodies and structural heart anomalies [4]. However, CHB associated with structural abnormalities or foetal hydrops has been recognised as a major risk factor for both prenatal and postnatal mortality.

In one of the largest reported series, Lopes LM et al., documented that 13.7% of 116 neonates had non-immune-mediated CHB. Among these, 4.3% spontaneously reverted to sinus rhythm without any intervention, reinforcing the notion that spontaneous regression is possible, although the underlying mechanisms remain poorly understood [5].

A separate cohort study examining 40 foetal conduction abnormalities also included cases of isolated, seronegative AVB. Among these, three were managed with dexamethasone therapy during pregnancy. In one case, the AVB reverted to sinus rhythm and the associated hydrops resolved. The authors emphasised that "isolated seronegative AVB exhibits a favourable long-term prognosis, with the possibility of spontaneous regression if the mother remains seronegative throughout pregnancy" [6].

Chang YL et al., described spontaneous resolution in six of seven foetuses diagnosed with second-degree AVB, most of which demonstrated unstable conduction patterns. These fluctuations in AVB severity often returned to normal sinus rhythm either before or shortly after birth [7]. Similarly, Breur JM et al., reported that rhythm variability may serve as a distinguishing feature of transient, non-immune-mediated AVB [8].

In alignment with these observations, Vassileva Z et al., presented a case involving a structurally normal foetus with second-degree 2:1 AVB and negative maternal autoantibodies, in which spontaneous resolution occurred by 25 weeks' gestation. The authors highlighted the rarity of such presentations, noting that only a limited number of non-immune, self-resolving 2:1 foetal AVB cases have been documented in the literature. Their findings reinforce the value of a conservative, observation-based approach to management in antibody-negative pregnancies without structural heart anomalies [9].

In contrast, the case described in our study involved a persistent AVB that spontaneously resolved in utero at 23 weeks' gestation. Notably, this case lacked the fluctuating conduction patterns seen in earlier reports, making it distinct. Similarly, Kasar T et al., described postnatal spontaneous resolution of congenital complete heart block on day 15 of life in twins born to seronegative mothers, further illustrating the potential for delayed resolution [10].

When examining long-term outcomes, Berg C et al., observed similar mortality rates among children born to anti-Ro-positive and negative mothers, suggesting that maternal antibody status may not be the sole prognostic determinant [11]. Moreover, a large multicentre review by Baruteau AE et al., reported that while 79.4% of children with congenital or childhood-onset AVB required pacemaker implantation, 90.1% maintained preserved systolic function during long-term follow-up, indicating generally favourable functional outcomes with appropriate management [12].

It is crucial to interpret maternal anti-Ro/La negativity with caution. While negative serology reduces the likelihood of an immune-mediated mechanism, it does not definitively rule it out. Some studies report late seroconversion, and factors such as low antibody titres, assay sensitivity limitations, or unrecognised maternal-foetal immune interactions may account for such cases [4]. This raises the possibility of additional, unidentified maternal or foetal contributors to AVB pathogenesis.

In the absence of identifiable immune or structural causes, transient AVB may result from increased vagal tone, immature conduction pathways, or transient functional insults [13]. However, these explanations remain speculative, and further research is warranted to better understand the underlying mechanisms [14].

CONCLUSION(S)

This outcome highlights the potential for a benign clinical course in cases of isolated, non-progressive CHB without structural cardiac anomalies, particularly when spontaneous resolution occurs in utero. While the pathophysiology of complete AVB, its disease progression, and the mechanisms underlying spontaneous resolution remain poorly understood, this case underscores the need for further research. Specifically, multicentre prospective studies are essential to identify prenatal and perinatal risk factors, elucidate mechanisms of transient resolution, and evaluate the role of genetic mutations, polymorphisms, serum biomarkers, and echocardiographic parameters. These efforts would help predict the likelihood and timing of spontaneous resolution in non-immune-mediated CHB occurring in structurally normal hearts. Improved understanding in these areas would allow clinicians to provide more accurate counselling and prognostication to families, reduce parental anxiety, and potentially limit unnecessary interventions such as antenatal corticosteroid use and pacemaker therapy.

REFERENCES

[1] Campbell R, Fischbach P, Frias P, Strieper M. Congenital complete heart block. Eur Cardiovasc Dis. 2007;3(2):11. Doi: 10.15420/ecr.2007.0.2.111.

[2] Ju YT, Wei YJ, Hsieh ML, Wang JN, Wu JM. Transient congenital complete heart block: A case report. Children (Basel). 2021;8(9):790. Doi: 10.3390/children8090790.
[3] Shao S, Liao H, Zhou S. Isolated non-immune-mediated second-degree atrioventricular block in the foetus: Natural history and predictive factors for spontaneous recovery. Ultrasound Obstet Gynecol. 2024;64(4):486-92. Doi: 10.1002/uog.27662.
[4] Maeno Y, Himeno W, Saito A. Clinical course of fetal congenital atrioventricular block in the Japanese population: A multicentre experience. Heart. 2005;91(8):1075-79. Doi: 10.1136/hrt.2003.033407.
[5] Lopes LM, Tavares GM, Damiano AP. Perinatal outcome of fetal atrioventricular block: One hundred sixteen cases from a single institution. Circulation. 2008;118(12):1268-75. (Published correction appears in Circulation. 2008 Oct 14;118(16):e671.) Doi: 10.1161/CIRCULATIONAHA.107.735118.
[6] Yadav M, Gowda M. Fetal conduction abnormalities: Retrospective analysis of 40 cases from a South Indian tertiary care centre with management insights and brief review. Prenatal Cardiol. 2024;14(1). Doi: 10.5114/pcard.2024.142415.
[7] Chang YL, Hsieh PC, Chang SD, Chao AS, Liang CC, Soong YK. Perinatal outcome of fetus with isolated congenital second degree atrioventricular block without maternal anti SSA/Ro SSB/La antibodies. Eur J Obstet Gynecol Reprod Biol. 2005;122(2):167-71. Doi: 10.1016/j.ejogrb.2005.01.013.
[8] Breur JM, Oudijk MA, Stoutenbeek P, Visser GH, Meijboom EJ. Transient non autoimmune fetal heart block. Fetal Diagn Ther. 2005;20(2):81-85. Doi: 10.1159/000082427.
[9] Vassileva Z, Sucu M, Aydin K, Aydin A. Transient fetal non-immune second-degree 2:1 atrioventricular block: A case report. Turk J Womens Health Neonatol. 2024;6(3):100-04.
[10] Kasar T, Saygı M, Özyılmaz İ, Ergül Y. Spontaneous remission of congenital complete atrioventricular block in anti Ro/La antibody negative monozygotic twins: Case report. Balkan Med J. 2017;34(1):71-73. Doi: 10.4274/balkanmedj.2015.0535.
[11] Berg C, Geipel A, Kohl T. Atrioventricular block detected in fetal life: Associated anomalies and potential prognostic markers. Ultrasound Obstet Gynecol. 2005;26(1):4-15. Doi: 10.1002/uog.1918.
[12] Baruteau AE, Pass RH, Thambo JB. Congenital and childhood atrioventricular blocks: Pathophysiology and contemporary management. Eur J Pediatr. 2016;175(9):1235-48. Doi: 10.1007/s00431 016 2748 0.
[13] Kikano SD, Killen SAS. Transient fetal atrioventricular block: A series of four cases and approach to management. J Cardiovasc Electrophysiol. 2022;33(10):2228-32. Doi: 10.1111/jce.15642.
[14] De Carolis S, Garufi C, Garufi E, De Carolis MP, Botta A, et al. Autoimmune congenital heart block: A review of biomarkers and management of pregnancy. Front Pediatr. 2020;8:607515. Doi: 10.3389/fped.2020.607515.

PARTICULARS OF CONTRIBUTORS:
1. Senior Resident, Department of Paediatrics, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India.
2. Senior Resident, Department of Paediatrics, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Sree Divya Mohan,
7, CLC Works Road, Shankar Nagar, Chromepet, Chennai-600044,
Tamil Nadu, India.
E-mail: drsreedivyamohan@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Feb 27, 2025
- Manual Googling: Jul 11, 2025
- iThenticate Software: Jul 14, 2025 (6%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Feb 26, 2025**
Date of Peer Review: **Jun 18, 2025**
Date of Acceptance: **Jul 17, 2025**
Date of Publishing: **Oct 01, 2025**