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Case Report

Congenital complete heart block in a neonate-not always maternal lupus

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ABSTRACT

Congenital complete heart block (CCHB) is a rare cardiac disorder in neonates and maternal lupus is the most common cause. More rarely, CCHB can be associated with congenitally corrected transposition of the great arteries. In this case, the neonate was born via emergency caesarean section due to bradycardia. Post-natal electrocardiogram and echocardiography confirmed the diagnosis. A pacemaker was inserted and the baby was kept under follow-up.

Keywords: Complete congenital heart block, Transposition of great arteries, Pacemaker

INTRODUCTION

Congenital complete heart block (CCHB) is a rare cardiac disorder with an estimated incidence of 1 per 15,000 to 20,000 live births.[1] CCHB is diagnosed in utero, at birth/ within 1st month of life. Commonly, CCHB occurs with maternal lupus due to transplacental passage of anti-Ro and/or anti-La antibodies to the fetus. More rarely, CCHB is associated with congenital heart disease (CHD), like congenitally corrected transposition of great arteries (CCTGA) with incidence of 1 per 33000 live births (accounting for 0.05% of CHD).[2] CCTGA is also described as "double discordance" characterized by discordant connections at both AV and VA junctions resulting in normal physiology.[3] Here, we present a rare case of CCHB with CCTGA managed successfully.

CASE REPORT

A female neonate was born to 32-year-old, gravida 3 para 1 abortus 1 mother at gestational age of 34 weeks. In current pregnancy, mother was diagnosed to have diabetes mellitus in 1st trimester and started on metformin followed by insulin. Mother also had hypothyroidism (was on thyroxine). No unsupervised medications were taken in current pregnancy. There was no history suggestive of autoimmune disease in mother. Anti-Ro and Anti-La workup was also negative. In previous pregnancy also, she had diabetes and hypothyroidism. But as per mother, in inter-pregnancy period, she didn't take any medication for same. She was referred to our institute as case of fetal bradycardia. Ultrasound exam also showed fetal bradycardia with heart rate of 78 bpm. Amniotic fluid was not meconium stained. Emergency cesarean section was done and baby cried immediately after birth with APGAR score 7 and 9 at 1 and 5 min of life. Birth weight was 2100 gm and baby are appropriate for gestational age. Baby was admitted to NICU and on admission heart rate was 70 bpm with normal blood pressure of 61/42 (49). Baby developed transient tachypnea of newborn after birth for which she was given on CPAP support. Chest X-ray revealed mild cardiomegaly (Figure 1 A). Electrocardiogram showed complete atrioventricular block (AV), with atrial rate of 166 bpm and ventricular rate of 75 bpm (Figure 2 A). ECHO done which was suggestive of CCTGA, with PFO of 5 mm, small PDA with L to R shunt (Figure 3). Heart rate varied between 55-70 bpm. On 4th day of life,

permanent VVIR mode pacemaker was implanted in epicardium left ventricle.[4] Post pacemaker implantation (Figure 1 B) heart rate was 120 bpm (Figure 2 B) with stable vitals. No syndromic features noted. Cranial, abdominal and renal ultrasounds normal. Patient was discharged successfully and kept under follow-up.

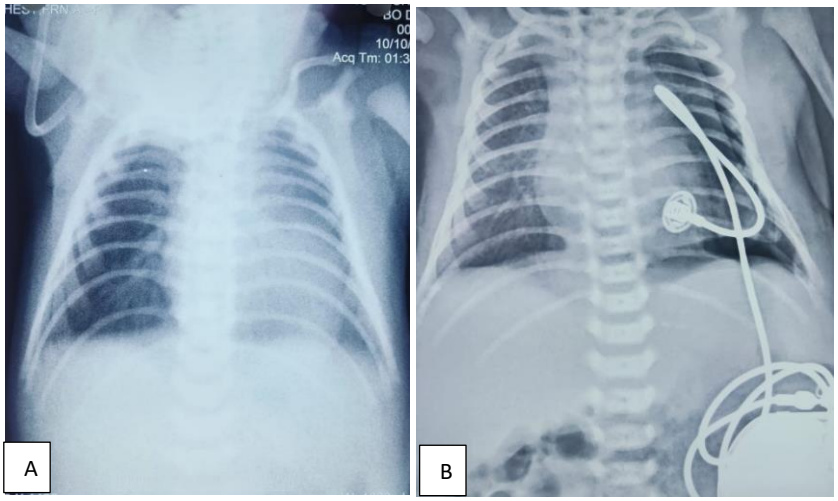

Figure 1 (A and B): Chest x-ray showing mild cardiomegaly, X-ray after pacemaker implantation.

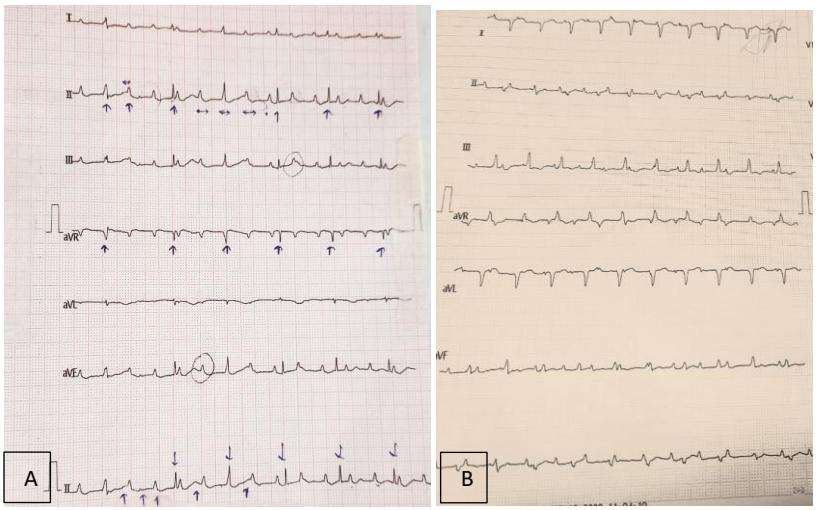

Figure 2 (A and B): ECG showed complete heart block with ventricular rate of 75 bpm and post pacemaker insertion ECG showed normal heart rate.

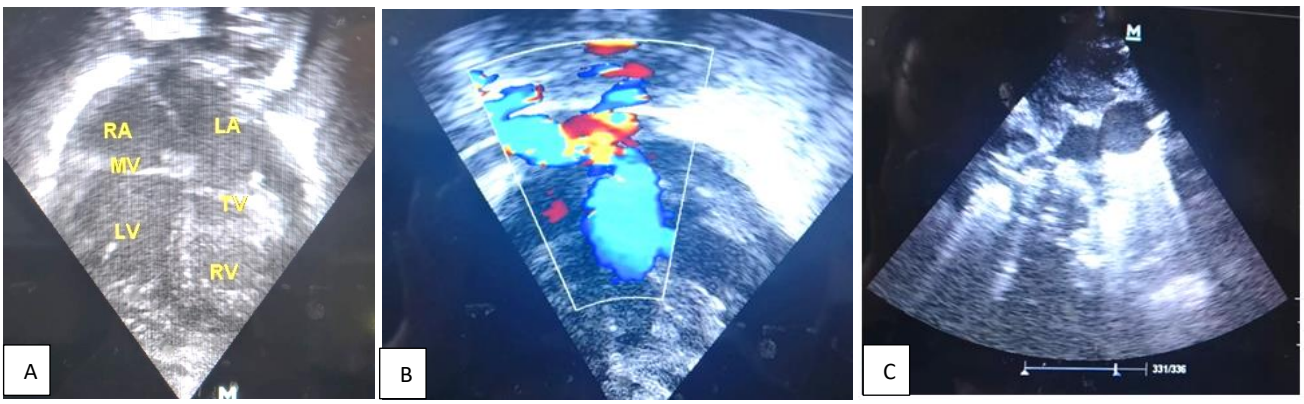

Figure 3 (A-C): Echocardiography showed CCTGA: showing 4 chamber view, with the right AV valve insertion higher than the left AV valve, showing blood from LV flowing to the pulmonary artery, short axis view showing aorta is present to the left and to anterior to the pulmonary artery.

DISCUSSION

CCTGA is a rare CHD which may be associated with cardiac rhythm abnormalities due to abnormal AV-node and a bundle of his.[5,6] Because of limited resources, no fetal echocardiography was done and fetal bradycardia was detected in the last trimester in our patient. Poorly controlled maternal diabetes mellitus increases the risk of TGA in fetus which was also seen in our case.[7] CCTGA is associated with structural heart defects like VSD (70-80%), pulmonary outflow tract obstruction (30-60%), aberrant mitral valve, and tricuspid abnormalities.[3,5] CCTGA is linked to arrhythmias, the most prevalent of which is CHB. The SA node is located normally; however, cardiac conduction abnormalities are common because of the AV node's aberrant position and the AV bundle's path.[8-10] Even though CCTGA may be detected in the prenatal stage by ultrasound, fetal echocardiography, and MRI, it may go undiscovered in countries with limited resources. A four-chamber view can be used to diagnose it in the antenatal period. In echocardiography, it is advised to concentrate on the distinction of the left and right ventricles. In particular, the morphologic right ventricle may be recognised in the four-chamber view due to its posterior and left position, its prominent moderator band, its more irregular endocardial surface, its more apical attachment of the atrioventricular (tricuspid) valve, and its distal and central attachment of the papillary muscles.[11,13] Approximately 10% of CCTGA patients have isolated presentation, and initially, they often exhibit no symptoms. However, by the time they reach their fourth or fifth decade of life, these individuals may start to have symptoms because of progressive TR, congestive heart failure, heart block, or ventricular arrhythmia.[14] In our case, CCTGA was associated with CHB (congenital heart block) which was confirmed via electrocardiogram and echocardiography. In our patient, heart rate was ranging between 55- 70 bpm, with structural heart disease of CCTGA, with PDA and PFO, so according to indication cardiac pacemaker insertion was planned.[15,16] In our patient VVIR mode, an epicardial pacemaker with lead in the left ventricle is inserted.[4] It is now recognized that a subset of paced patients develops dilated cardiomyopathy and heart failure, therefore long-term regular follow-up is mandatory.[17]

CONCLUSION

CCTGA with CHB is a complex heart disease which requires a multidisciplinary approach for management. Good antenatal follow-up and delivery at a tertiary care institute are key for good outcomes. Long-term follow-up is required in these patients for early identification of heart failure.

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REFERENCES

1. Nkoke C, Wawo EY, Mfeukeu LK, Makamte L, Edie SD, Balana FE. Complete congenital heart block in a neonate with a complex congenital heart defect in Africa. Cardiovasc Diagn Ther. 2016;6(1):S78-82.
2. Van der Linde D, Konings EEM, Slager MA. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. J Am Coll Cardiol. 2011;58(21):2241-7.
3. Wallis GA, Debich-Spicer D, Anderson RH. Congenitally corrected transposition. Orphanet J Rare Dis. 2011;6(1):22.
4. 2002 NASPE Position Statement: The Revised NASPE/BPEG Generic Code for Antibradycardia, Adaptive-Rate, and Multisite Pacing. Heart Rhythm Society. Available at: https://www.hrsonline.org/guidance/clinical-resources/2002-naspe-position-statement-revised-naspebpeg-generic-code-antibradycardia-adaptive-rate-and. Accessed on 23 November, 2023.
5. Baruteau A, Abrams DJ, Ho SY, Thambo J, McLeod CJ, Shah MJ. Cardiac Conduction System in Congenitally Corrected Transposition of the Great Arteries and Its Clinical Relevance. JAHA. 2017;6(12):e007759.
6. Anderson RH, Becker AE, Arnold R, Wilkinson JL. The conducting tissues in congenitally corrected transposition. Circulation. 1974;50(5):911-23.
7. Alfarhan A, Alquayt M, Alshalhoub M, Alnahdi MA, Masuadi E, Alhabshan F. Risk factors for transposition of the great arteries in Saudi population. Saudi Med J. 2020;41(10):1054-62.
8. Huhta JC, Maloney JD, Ritter DG, Ilstrup DM, Feldt RH. Complete atrioventricular block in patients with atrioventricular discordance. Circulation. 1983;67(6):1374-77.
9. Daliento L, Corrado D, Buja G, John N, Nava A, Thiene G. Rhythm and conduction disturbances in isolated, congenitally corrected transposition of the great arteries. Am J Cardiol. 1986;58(3):314-8.
10. Gillette PC, Busch U, Mullins CE, McNamara DG. Electrophysiologic studies in patients with ventricular inversion and “corrected transposition”. Circulation. 1979;60(4):939-45.
11. Paladini D, Volpe P, Marasini M, Russo MG, Vassalo M, Gentile M et al. Diagnosis, characterization and outcome of congenitally corrected transposition of the great arteries in the fetus: a multicenter series of 30 cases. Ultrasound Obstet Gynecol. 2006;27(3):281-5.
12. Sharland G, Tingay R, Jones A, Simpson J. Atrioventricular and ventriculoarterial discordance (congenitally corrected transposition of the great arteries): echocardiographic features, associations, and outcome in 34 fetuses. Heart. 2005;91(11):1453-8.
13. McEwing RL, Chaoui R. Congenitally corrected transposition of the great arteries: clues for prenatal

diagnosis. Ultrasound Obstet Gynecol. 2004;23(1):68-72.
14. Presbitero P, Somerville J, Rabajoli F, Stone S, Conte MR. Corrected transposition of the great arteries without associated defects in adult patients: clinical profile and follow up. Br Heart J. 1995;74(1):57-59.
15. Khairy P, Hare GFV, Balaji S, Berul CI, Cecchin F, Cohen MI et al. PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease: Developed in Partnership Between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the Governing Bodies of PACES, HRS, the American College of Cardiology (ACC), the American Heart Association (AHA), the European Heart Rhythm Association (EHRA), the Canadian Heart Rhythm Society (CHRS), and the International Society for Adult Congenital Heart Disease (ISACHD). Can J Cardiol. 2014;30(10):e1-63.
16. Brignole M, Auricchio A, Baron-Esquivias G, Bordachar P, Boriani G, Breithardt OA et al. 2013 ESC Guidelines on cardiac pacing and cardiac resynchronization therapy: the Task Force on cardiac pacing and resynchronization therapy of the European Society of Cardiology (ESC). Developed in collaboration with the European Heart Rhythm Association (EHRA). Eur Heart J. 2013;34(29):2281-29.
17. Benrey J, Gillette PC, Nasrallah AT, Hallman GL. Permanent pacemaker implantation in infants, children, and adolescents. Long-term follow-up. Circulation. 1976;53(2):245-48.

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