

Complete Congenital Heart Block in a Neonatal Lupus Erythematosus Associated with Pulmonary Involvement without Pacemaker Implantation: A Case Report

Yazdan Ghandi^{1*}, Hamideh Kazemi², Saeed Alinejad³, Mehrzad Sharifi⁴

1. Assistant Professor, Pediatric Cardiologist, Amirkabir Hospital, Arak University of Medical Sciences, Arak, Iran

2. Pediatric Resident, Amirkabir Hospital, Arak University of Medical Sciences, Arak, Iran

3. Assistant Professor, Neonatologist, Amirkabir Hospital, Arak University of Medical Sciences, Arak, Iran

4. Assistant Professor, Amiralmomenin Hospital, Department of Cardiac Surgery, School of Medicine, Arak University of Medical Sciences, Arak, Iran

ABSTRACT

Background: Neonatal lupus erythematosus is an uncommon disease. Congenital complete heart block (CCHB) usually happens in neonates with maternal systemic lupus erythematosus. The most prevalent presentation of CCHB is bradycardia that can be diagnosed through an electrocardiogram.

Case report: Here in, we present the case of a full-term male neonate with gestational age of 37 weeks and birth weight of 2200 g, whose mother had positive anti-Ro/SSA antibodies. The mother was asymptomatic without any criteria of systemic lupus erythematosus. The newborn presented with bradycardia, respiratory distress and narrow QRS complex without structural heart disease. He was connected to mechanical ventilator and did not need pacemaker implantation.

Conclusion: This case report was conducted on a newborn with CCHB associated with pulmonary disorder. The newborn was intubated due to respiratory distress and did not need pacemaker implantation; however, after 8-month follow up, excellent outcomes were observed. It seems that atelectasis and mechanical ventilation can intolerance, and pacemaker implantation did not need in NLE with CCHB with narrowing QRS complex.

Keywords: Anti-Ro/SSA autoantibodies, Arrhythmia, Bradycardia, Congenital complete heart block, Neonatal lupus erythematosus

Introduction

The congenital complete heart block (CCHB) is a rare disorder with high morbidity and mortality rates. The incidence rate of this disease is reported to range between 1: 15,000 and 1: 22,000 live births (1). CCHB is associated with maternal lupus or other autoimmune diseases such as Jorgen's syndrome (2). CCHB happens with the transplacental passage of maternal anti-Ro/SSA and anti-La/SSB autoantibodies, which affects atrioventricular node of fetus and damages the development of cardiac conduction system (3). The total incidence rate of fetal CCHB through anti-Ro/La-positive mothers is reported to be about 1-5%. However, the risk of CCHB occurrence rises up to 25% for those with previous affected children. Moreover, the risk of CCHB is higher if the mother has a previous child with neonatal lupus erythematosus (NLE) or hypothyroidism due to autoantibodies. The most common clinical manifestations of NLE are related to cutaneous, cardiac, and hepatobiliary abnormalities. Some infants also present with

hematologic disorders such as pancytopenia. The co-occurrence of CCHB with structural cardiac abnormalities increases the rate of mortality (4, 5). Features of congenital atrioventricular block particularly depend on the timing of the presentation and the accompanying structural heart disease. Mothers can be absolutely asymptomatic with positive anti-Ro and anti-La autoantibodies. In this case, incidental findings such as bradycardia or hydrops fetalis are the only clues for further evaluations and diagnosis (6). Some evidence suggests that administration of steroids and immunoglobulins or plasmapheresis in mothers can reverse the complete heart block in the fetus (7). In the current study, we present the case of a newborn with isolated CCHB without congenital cardiac disease or other NLE complications. This infant did not require pacemaker implantation.

* Corresponding author: Yazdan Ghandi, Arak University of Medical Sciences, No. 62 Alamolhoda Street, Amirkabir Hospital, Iran. Tel: +988633135075; Email: drghandi_1351@yahoo.com

Case report

A 1-day-old full-term male neonate with birth weight of 2200 g, admitted to Neonatal Intensive Care Unit (NICU) due to bradycardia (60-70 bpm) and respiratory distress. His mother was 22 years old (gravid: 2, para: 1, abortus: 0) with normal examine and normal previous delivery. In 29 weeks of gestation, she was referred to the emergency department because of fetal bradycardia (70-80 bpm), diagnosed through a routine abdominal ultrasound examination. Ultrasound revealed no evidence of fetal distress. Fetal echocardiography showed CCHB without any structural heart damage (isolated congenital heart block). She did not present any symptoms of autoimmune disease; however, immunological tests showed positive anti-Ro/SSA (titer: 37.5). Antibody Ro/SSA was detected by enzyme-linked immune-osorbent assay (ELISA) and Western blot immune-odiffusion tests.

She was offered to receive dexamethasone, but she did not accept any medication such as intravenous immunoglobulin or corticosteroids therapy. She was followed up by weekly ultrasound tests for the evaluation of the fetal heart rate, which was persistent (60-65 bpm), and no signs of hydrops fetalis were observed. At 37 weeks of gestation, caesarean section was performed due to pre-eclampsia; the heart rate was 65 bpm at birth (Figure 1). At NICU, the newborn was intubated due to respiratory distress and extubated after about 48 h; at this time, he showed the stable heart rate of 65 bpm.

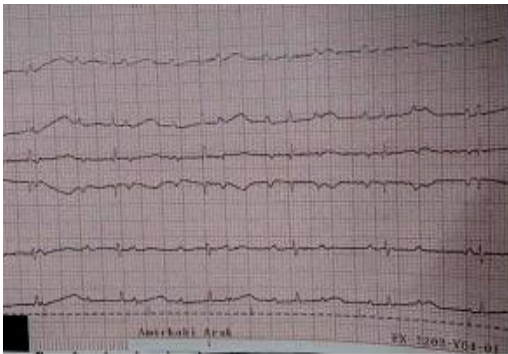


Figure 1. Post-delivery echocardiography showed complete congenital heart block

Biochemical tests and arterial blood gas evaluation were within the normal range. Echocardiography revealed no cardiac structural abnormalities such as congenitally corrected transposition of the great arteries, Ebstein's anomaly, left isomerism resulted in cardiac malfunctions, and left ventricular non-compaction. Left ventricular size

and systolic function were normal. The neonate did not have heart failure. There was no feature of cardiomegaly on chest x-ray (CXR), but there was atelectasis (a collapse of the top of the right lung) that was entirely resolved with treatment in the next graph (Figures 2 a, b).

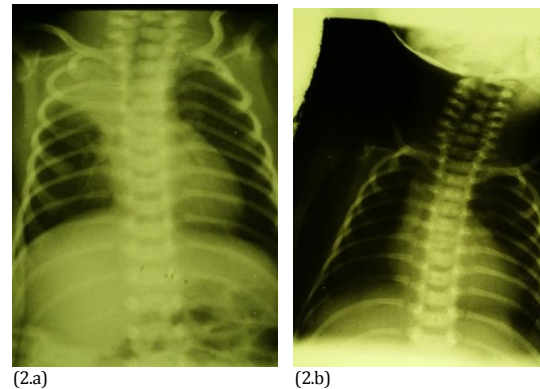


Figure 2 (a, b). Anteroposterior chest X-ray view showing normal heart size (note the right lung collapse [atelectasis] before and after treatment)

The aspiration pneumonia of the right lung could be caused by breastfeeding on the first day. The neonate was carefully followed up for 10 days, and he did not require pacemaker implantation. In the follow up visit, the neonate underwent heart monitoring for 48 h, during which the average heart rate was not lower than 60 bpm. During 10 days of hospital stay, daily echocardiography showed a wide QRS; however, the newborn had good general condition and was breast-fed. The wide QRS is a risk factor for sudden death. After an eight-month follow up, the QRS was no longer wide.

ECG demonstrates no relation between P wave and QRS complex. The P wave had a normal shape; the QRS complex was not wide.

Discussion

NLE is an uncommon transplacentally acquired autoimmune disorder, which affects atrioventricular node and His bundles of fetal heart and can cause cardiac fibrosis and calcification (3). Fetal exposure to these maternal autoantibodies can cause myocarditis and late-onset cardiomyopathy (8). Our patient suffered from isolated CCHB without any evidence of structural damage or myocarditis. The peak onset of the diagnosis of CCHB is between 18 and 24 weeks of gestation, about six weeks after effective placental transport of maternal IgG antibodies. The passage of maternal autoantibodies causes fetal bradycardia, which is detected by fetal echocardiography. In the patient, bradycardia was diagnosed at 29 weeks of gestation through a routine ultrasound test; however, due to the mother's

disinclination, the clinical protocol was not administered. Jaeggi and colleagues reviewed 102 patients (aged over 30 years) with CCHB (9). They reported that patients diagnosed with CCHB in utero, after birth, and in childhood had mortality rates of 43%, 6%, and 0 %, respectively, within the first two decades of life. Hydrops fetalis, endocardial fibroelastosis, and delivery at less than 32 weeks increase the rate of mortality in patients with CCHB (9). Despite other manifestations of lupus erythematosus syndrome, heart block is not reversible. The degree of heart block in NLE may vary from first degree to third degree block, but most cases diagnosed in utero present with at least second degree or more advanced block. Approximately two-thirds of these patients need pacemaker (1).

The occurrence of early death in the majority of CCHB patients is due to delayed implantation of pacemaker or hemodynamic instability associated with significant congenital heart disease (10). Indications for pacemaker therapy are symptomatic bradycardia or heart rate <55 bpm in patients without structural abnormality. These indications in patients with structural damage correspond to heart rate <70 bpm, cardiomyopathy, and ventricular dysfunction (1). Our case showed no need for pacemaker during the eight-month follow up.

Dexamethasone is one of the treatment options during pregnancy since it is not metabolized by the placenta. Plasmapheresis and beta-mimetic agents are other treatments for this disorder (11). However, in the current study, no treatment was performed during pregnancy due to the mother's disagreement. In a case series study evaluating 16 patients with isolated CCHB and late-onset cardiomyopathy, the occurrence of late-onset cardiomyopathy was reported to be 5-11% despite the implantation of pacemaker in the CCHB patients (10). Therefore, it is recommended to the infants' families to have regular and precise heart monitoring not only in terms of rhythm and rate, but also for ventricular function and late-onset cardiomyopathy.

Conclusion

It seems that atelectasis and mechanical ventilation intolerance, and pacemaker implantation did not need in NLE with CCHB with narrowing QRS complex. We successfully managed a case of CCHB without inserting temporary pacemaker; however, the patient was intubated due to respiratory distress and atelectasis. Early detection and timely management are the keys to success in the treatment of newborns.

Acknowledgments

None.

Conflicts of interests

None declared.

References

1. Friedman DM, Duncanson LJ, Glickstein J, Buyon JP. A review of congenital heart block. *Images Paediatr Cardiol.* 2003; 5(3):36-48.
2. Claus R, Hickstein H, Kulz T, Lenschow U, Meiske D, Kotitschke A, et al. Identification and management of fetuses at risk for, or affected by, congenital heart block associated with autoantibodies to SSA (Ro), SSB (La), or a HsEg5-like autoantigen. *Rheumatol Int.* 2006; 26(10):886-95.
3. Jaeggi E, Laskin C, Hamilton R, Silverman E. The importance of maternal anti-Ro/SSA antibodies as prognostic markers of the development of cardiac neonatal lupus erythematosus: a prospective study of 186 antibody-exposed fetuses and infants. *J Am Coll Cardiol.* 2010; 55(24):2778-84.
4. Costedoat-Chalumeau N, Amoura Z, Lupoglazoff JM, Huang DL, Denjoy I, Vauthier D, et al. Outcome of pregnancies in patients with anti-SSA/Ro antibodies: a study of 165 pregnancies, with special focus on electrocardiographic variations in the children and comparison with a control group. *Arthritis Rheum.* 2004; 50(10):3187-94.
5. Brucato A, Frassi M, Franceschini F, Cimaz R, Faden D, Pison MP, et al. A Risk of congenital complete heart block in newborns of mothers with anti-Ro/SSA antibodies detected by counterimmunoelectrophoresis: a prospective study of 100 women. *Arthritis Rheum.* 2001; 44(8):1832-5.
6. Hutter D, Silverman ED, Jaeggi ET. The benefits of transplacental treatment of isolated congenital complete heart block associated with maternal anti-Ro/SSA antibodies: a review. *Scand J Immunol.* 2010; 72(3):235-41.
7. Friedman DM, Kim MY, Copel JA, Davis C, Phoon CK, Glickstein JS, et al. Utility of cardiac monitoring in fetuses at risk for congenital heart block: the PR Interval and Dexamethasone Evaluation (PRIDE) prospective study. *Circulation.* 2008; 117(4):485-93.
8. Ayed K, Gorgi Y, Sfar I, Khrouf M. Congenital heart block associated with maternal anti SSA/SSB antibodies: a report of four cases. *Pathol Biol (Paris).* 2004; 52(3):138-47.
9. Jaeggi ET, Hamilton RM, Silverman ED, Zamora SA, Hornberger LK. Outcome of children with fetal, neonatal or childhood diagnosis of isolated congenital atrioventricular block. A single institution's experience of 30 years. *J Am Coll Cardiol.* 2002; 39(1):130-7.
10. Moak JP, Barron KS, Hougen TJ, Wiles HB, Balaji S, Sreeram N, et al. Congenital heart block: development of late-onset cardiomyopathy, a previously underappreciated sequela. *J Am Coll Cardiol.* 2001; 37(1):238-42.

11. Friedman DM, Rupel A, Glickstein J, Buyon JP. Congenital heart block in neonatal lupus: The

pediatric cardiologist's perspective. *Indian J Pediatr.* 2002; 69(6):517-22.