Original Article

Rare Case of Congenital Heart Block in a Retrospectively Diagnosed Case of Sjögren's Syndrome in Mother: A Case Report

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Abstract

Sjögren's (SHOW'-grenz) syndrome is a systemic autoimmune, rheumatic disease that affects the entire body. The most common problems are dry mouth, dry eyes, fatigue and musculoskeletal pain in adults. Ten times as many women as men are diagnosed with Sjögren's. While most often diagnosed in women during middle age or after menopause. The congenital heart block (CHB) is defined as the heart block that is diagnosed in fetus (in utero) or within the first 28 days after birth (neonatal period). Congenital heart block is a rare disorder that appears to affect males and females in equal numbers. The most common cause of congenital heart block (CHB) is neonatal lupus due to maternal Sjögren's syndrome, an acquired autoimmune disease caused by transplacental transfer of maternal antibodies to the fetus. Several studies have reported an increased rate of spontaneous abortion and fetal loss associated with Sjögren syndrome. Congenital heart block occurs in a frequency of 1 in 20,000 live births. It has been reported to occur in 2% of Ro-positive mothers [2]; 5% of mothers with a diagnosis of mixed connective tissue and/or Sjögren Syndrome [3] and in 8% of Ro-positive mothers. Here we present an early pre-term neonate that was admitted to neonatal intensive care unit for bradycardia with stable haemodynamics. The mother, who showed no clinical symptoms or any particular history, was transferred to our tertiary centre for profound fetal bradycardia on recent scans. At birth, the infant's ECG showed a third-degree atrioventricular block and echocardiography was normal. Cardiac neonatal lupus was confirmed with positive maternal anti-Ro antibodies. Under close monitoring, the infant tolerated the bradycardia well (median 72 beats per minute (bpm)) and was discharged on day 21 of life. There was no indication for pacemaker at discharge, but he would be on regular follow-up with a paediatric cardiologist.

Keywords: AV node; Anti-La/SSB antibodies; Anti Ro/SSA antibodies; Atrioventricular block; Congenital heart block; Atrioventricular block; Neonatal lupus; Sjogren's syndrome; Echocardiography; Electrocardiogram.

INTRODUCTION

Atrioventricular block (AVB), also known as congenital heart block (CHB), is defined by interference with the electrical nerve impulses' conduction, which controls the heart's normal and rhythmic pumping function. Subdivisions of Congenital Heart Block are; first degree congenital heart block, second degree congenital heart block [Mobitz type I (Wenckebach); Mobitz type II] and third degree congenital (complete) heart block. Each new born impacted by such conduction anomalies has a different level of severity. In new-borns affected by CHB, the primary finding is a slow heart rate (bradycardia). Pregnant Sjogren's patients who are positive for the autoantibody SSA/Ro and SSA/La should seek care from a perinatologist (obstetrician who manages high risk pregnancies) and be monitored for signs of foetal heart block.[1-3]

The resultant fibrosis of foetal AV node is thought to be due to immune mediated tissue damage to foetal heart following transplacental passage of maternal IgG autoantibodies, may also lead to arrhythmias in the foetal conduction system.[4,5] The prenatal diagnosis of congenital heart block is more common as cardiac imaging techniques are improving. A growing number of autoimmune cases are being diagnosed between 18 and 24 weeks of pregnancy, leading to a better prognosis. Diagnosis depends on the results of one or more cardiac imaging tests such as fetal electrocardiography (ECG) and fetal echocardiography. This may help determine the type of heart block and may rule out any structural heart anomalies.[6]

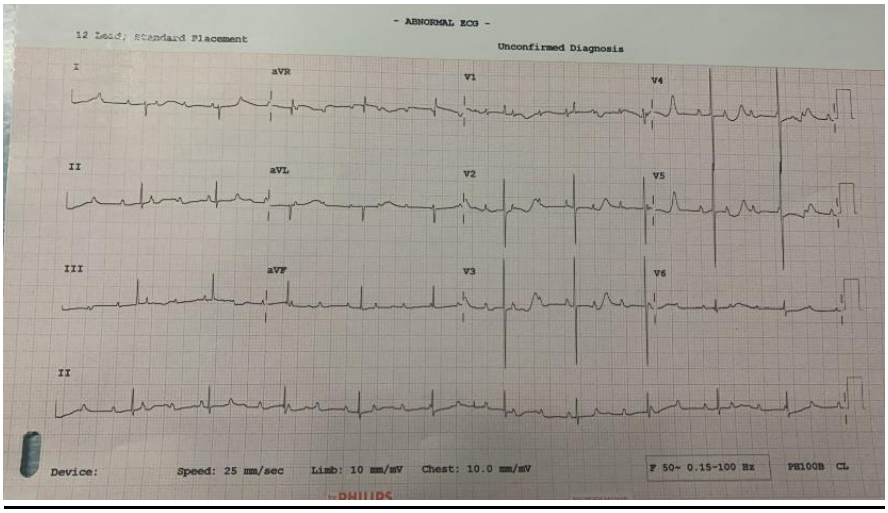

Figure 1: Electrocardiogram of the neonate showing bradycardia and (Lead II) QRS complexes being conducted at their own rate and totally independent of the P waves suggestive of a complete heart (atrioventricular) block.

Case report

A 24-year-old Primigravida retrospectively diagnosed with Sjogren's Syndrome (Anti Ro/SSA and Anti La/SSB positive titres found on further evaluation) presented at 34 weeks of gestation with complaints of persistent foetal bradycardia and oligohydramnios in antenatal scans. Foetal ECHO was suggestive of mild cardiomegaly with bradycardia, mild hypertrophy of both ventricles and atrioventricular block. Baby was delivered via LSCS, expected 32 weeks of neuromuscular and physical maturity by New Ballard's score, weighed 1.4kg, cried immediately after birth and routine neonatal care and resuscitation were followed. Immediately post-delivery aggressive cardiopulmonary resuscitation and use of adrenaline was avoided. Vitals at birth were as following, heart rate 60bpm, respiratory rate 70cpm, with significant distress, spO2 on room air 75%, peripheral and central pulses were well felt and a mean blood pressure of 48mmHg. Baby was intubated and taken on ventilator, surfactant was administered in view of severe hyaline membrane disease and restricted intravenous fluids with parenteral nutrition were added. Baby was weaned off to high flow nasal cannula, then room air. Throughout the 3-week course of admission, heart rate of the baby remained between 60-100 bpm. There was no indication for pacemaker at discharge, but he would be on regular follow-up with a paediatric cardiologist.

Figure 1 shows Electrocardiogram of the neonate showing bradycardia and QRS complexes being conducted at their own rate and totally independent of the P waves (lead II) suggestive of a complete heart (atrioventricular) block. Anti SSA/ SSB, Anti Ro-52 titres were found to be strongly positive in the baby. Baby was closely followed up and continues to feed well and gain weight (with heart rate still below 100bpm) at 2 months of age.

Discussion

In a normally conducting heart, sinoatrial node allows for atrial contraction, the atrioventricular node allows for transmission of the signal between the atria and ventricles, and the bundle of his-Purkinje system allows for ventricular contraction. As long as the electrical impulse is transmitted normally, the heart behaves and contracts normally allowing for blood to be pumped out to the body. When the transmission of the signal is impeded, the blocked electrical transmission is known as a heart block or an atrioventricular block. Congenital heart block occurs in a frequency of 1 in 20,000 live births.

First, second, or third-degree congenital heart block may be diagnosed between 18 and 24 weeks of gestation (complete). Mortality is close to 20%, and the majority of children who survive need pacemakers. Infants that are affected may develop cardiomyopathy. Anti-Ro/La antibodies are also connected to anomalies in new-borns' skin, liver, and blood.

On antenatal scan visits if fetal bradycardia is identified consistently, a 2-dimensional and M-mode fetal echocardiography and Doppler ultrasound should be obtained to determine whether there is an atrial arrhythmia or atrioventricular (AV) block, and to what degree, and whether there are major structural abnormalities of the heart. In such a case mother's serum should be tested by ELISA for anti-Ro and/or anti-La antibodies. [6,7]

According to a study the authors concluded that they were unable to detect first-degree AVB before progression to complete heart block on just fetal ECHO. Following which it was noted in 70 fetuses of 56 mothers that using tissue velocity fetal kinetocardiogram helped in measurement of PR prolongation which aided in an earlier diagnosis and prompt maternal treatment. Thus, Echo Doppler seems a less reliable method for early detection of fetus first-degree AVB, and it is suggested that fetal kinetocardiogram or fetal electrocardiography are preferred. Fluorinated steroids given on detection were associated with normalized atrioventricular conduction in foetuses with first-degree AVB. [8]

Thus, to summarize the possible treatment options for congenital heart block in pregnancy are few, yet foetuses frequently tolerate slow escape rhythms. Adrenocorticosteroids like dexamethasone, which are not metabolised by the placenta, may be provided to the mother if a more severe degree of heart block is present.

Dexamethasone works to decrease inflammation and the number of circulating maternal antibodies in the fetus. In affected individuals who exhibit mild forms of heart block (such as first degree or second degree Mobitz I), treatment may not be required. More severe cases (such as second degree Mobitz II- and third-degree heart block) may require a temporary or permanent pacemaker.[8] Presence of bradycardia (slow heart rate), congestive heart failure and structural heart disease are usually poor prognostic markers.

Conclusion

The most common cause of congenital heart block (CHB) is neonatal lupus, an acquired autoimmune disease caused by transplacental transfer of maternal antibodies to the fetus. We add to the sparse literature documenting congenital heart block. Pregnant women in whom fetal bradycardia is persistently seen on antenatal scans should prompt the health care professional to further go for detailed fetal echocardiography and auto-immune blood work up in the mother.

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