International Journal of Advances in Medicine
Kamath V et al. Int J Adv Med. 2014 May;1(1):37-39
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: 10.5455/2349-3933.ijam20140508

Case Report

Fetal isolated congenital heart block associated with maternal anti-SSA/SSB antibodies

Vidyashri Kamath*, Mahesh Navada, Dhara, Prema D'Cunha

Department of Obstetrics and Gynaecology, Father Muller Medical College, Mangalore, Karnataka, India

Received: 5 April 2014
Accepted: 27 April 2014

***Correspondence:**
Dr. Vidyashri Kamath,
E-mail: drvidyashri@gmail.com

ABSTRACT

Isolated Congenital Heart Block (CHB) affects 1:15000-20000 live births. 30-50% of fetuses with CHB will have a structural anomaly. Congenital heart block detected in utero is strongly associated with maternal antibodies to SSA (Ro) and SSB (La). Their pathogenic role in the development of CHB has been established in several studies. The mothers of affected infants frequently had autoimmune disease (systemic lupus erythematosus, Sjögren's syndrome) or were entirely asymptomatic. We report a case of fetal isolated congenital heart block in an asymptomatic mother with anti-SSA/SSB antibodies.

Keywords: Fetal congenital heart, Block, fetal bradycardia, Autoimmune antibodies

INTRODUCTION

Isolated Congenital Heart Block (CHB) affects 1:15000-20000 live births.[1] 30-50% of fetuses with CHB will have a structural anomaly. Congenital heart block detected in utero is strongly associated with maternal antibodies to SSA (Ro) and SSB (La). Their pathogenic role in the development of CHB has been established in several studies. The mothers of affected infants frequently had autoimmune disease (systemic lupus erythematosus, Sjögren's syndrome) or were entirely asymptomatic.[2] We report a case of fetal isolated congenital heart block in an asymptomatic mother with anti-SSA/SSB antibodies.

CASE REPORT

A 23 year old primigravida hailing from Bantwal, Karnataka was referred to our institution at 38 weeks of gestation with persistent fetal bradycardia since 20 weeks of gestation for further management. Maternal assessment revealed positive ANA Anti Rho and Anti LA antibodies. There were no symptoms of lupus or any connective tissue disorders. On examination she was of average built and nutrition with BP of 140/90 mm of Hg. Ophthalmologic and dermatological examination was found to be normal. On abdominal examination uterus was term size, relaxed, cephalic presentation. Fetal heart rate was 52-56 bpm. Hb was 11.5 g/dl. Peripheral smear showed normocytic normochromic blood picture. Blood sugar levels, liver function tests, kidney function tests were within normal limits. ESR-130mm (1st hour), CRP-29.6 mg/dl, 24 hour urine protein was 206.1 mg. PT, APTT was within normal limits. ANA profile was strongly positive (+++) for SS-A (60 kDa) and Ro-52 autoantibodies. Anticardiolipin (IgG and IgM) antibodies were negative. USG examination showed a single live intrauterine fetus of 37 weeks gestation with adequate liquor, EFW of 2.97 kg and FHR of 49 bpm. Fetal echocardiography (ECHO) showed no apparent structural defect, FHR being 48-52 bpm suggestive of congenital heart block.

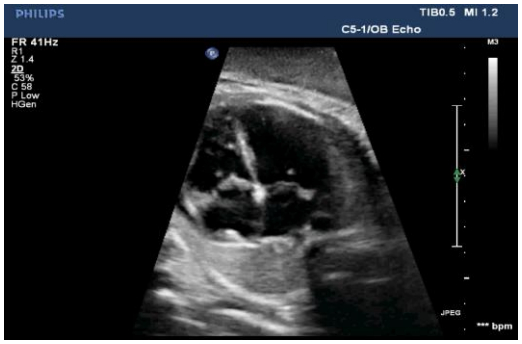

Figure 1: Fetal echo showing 4 chamber view.

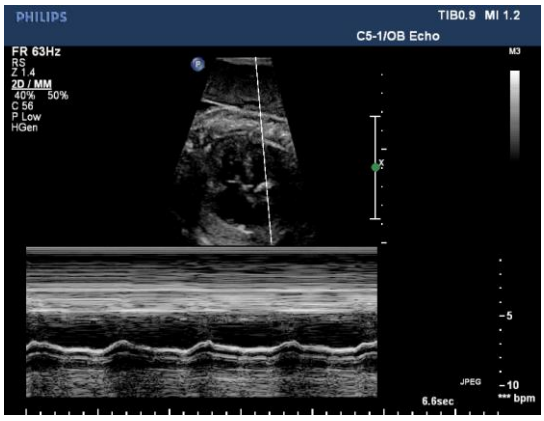

Figure 2: M mode identifying congenital heart block.

Maternal echocardiographic findings were normal. The couple was counseled regarding the prognosis of the fetus. Patient was started on tab. dexamethasone 4 mg OD. The following day of admission patient went into spontaneous labour. The couple was given the option of caesarean section because of the limitations in the interpretation of fetal heart monitoring. As the cardiovascular status of the fetus was stable and patient was already in labour, the couple wished for vaginal delivery and it was planned with careful monitoring of the fetus. Patient had a full term vaginal delivery of a live female baby 5 hours after the onset of labour. Birth weight was 3.12 kg with good APGAR. Baby was shifted to NICU for observation. HR was 52/min, RR was 38/min and oxygen saturation was maintained. ECG done showed complete heart block. ECHO showed ostium secundum ASD with mildly dilated left ventricle and good LV and RV function. She was started on tab. orciprenaline - 0.1 mg/kg TID. There were no features of neonatal lupus.

Our newborn had a ventricular rate of 50-55 beats/min, whereas normal newborns have a heart rate of 94 to 155 beats/min.[3] Despite her low heart rate, her weight and height were within normal limits. Signs of heart failure were not observed and no treatment was given during or after delivery. The parents were counseled about the need for pacemaker on further follow up. Mother and baby were discharged one week after birth.

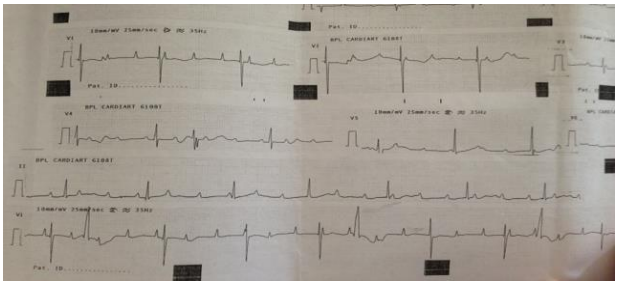

Figure 3: ECG of the newborn showing complete heart block.

DISCUSSION

CHB is a rare disorder and occurs in only 3% of infants born to mothers with anti-Ro/SSA and anti-La/SSB antibodies.[4] Antibodies to SSA/SSB have been proposed to be a serologic marker for neonatal lupus syndrome and CHB. CHB is presumed to be due to the transplacental passage of these IgG autoantibodies from the mother into the fetal circulation. It is caused by maternal anti-Ro and anti-La antibodies binding to cardiac tissue and causing a transient myocarditis and subsequent fibrosis of the conduction system at the atrioventricular node which is irreversible.[5] Fetuses with complete heart block have an increased mortality with >60% requiring permanent pacemakers and 10% developing severe cardiomyopathy. Most of the deaths occur in utero or during infancy. Premature birth, low birth weight, low ventricular rate, significant structural heart disease, evidence of ventricular dysfunction or associated cardiomyopathy, and the presence of hydrops fetalis are poor prognostic signs. Ascites and anasarca-type edema are also associated with poor outcome, and pacemaker implantation is indicated in infants with cardiac failure and a heart rate of under 55 beats/min. In the presence of hydrops fetalis, the reported mortality rates for infants born with CHB have exceeded 80%.[4,6,7] The risk of CHB increases in infants born to mothers with a previous child having CHB and occurs in nearly 18% of pregnancies subsequent to the index pregnancy with CHB.[4]

The outlook of patients with congenital heart block depends largely on the presence or absence of underlying structural heart disease, as well as the rate of ventricular activation and the presence or absence of congestive heart failure. Heart block with a normal cardiac structure is due to maternal anti-Ro or anti-La antibodies in the vast majority of cases. Prenatal therapy for such cases is controversial with some groups recommending therapy such as dexamethasone while others have tried IV immunoglobulins, plasmapheresis and azathioprine.[8] Transplacental treatment with dexamethasone can be considered for autoimmune mediated heart block, as it may prevent progressive heart block in fetuses with first or second degree block and prevent further damage to the myocardium in fetuses with third degree block.[8,9] If the cardiac structure is abnormal, the most common associated abnormalities include transposition of the great

vessels and atrioventricular septal defects. The prognosis for such fetuses, affected by both structural cardiac disease and complete heart block is guarded, with a minority of fetuses surviving.[10-13] Thereby frequent follow up in the antenatal period with twice weekly fetal echocardiography is necessary to ensure fetal wellbeing and to exclude the development of heart failure.

Mode of delivery

If persistent heart block is present during labor, some experts recommend a cesarean delivery because these arrhythmias limit the interpretation of fetal heart-rate monitoring. Others have suggested that fetal well-being can be assumed if there is variability in the ventricular heart-rate and an absence of decelerations, or with periodic biophysical profiles during labor. For this reason the management of labor and delivery in a fetus with heart block should be individualized based on discussion about the limitations of ensuring fetal wellbeing.

In conclusion, we believe that close fetal surveillance is needed for fetuses diagnosed with autoimmune CHB. Detail fetal echocardiography should be routinely performed. Delivery should be considered if there is evidence of fetal distress and/or deteriorating cardiac performance even in cases of prematurity. Screening of infants with isolated CHB or neonatal lupus and their mothers for the presence of anti-SSA and anti-SSB is strongly recommended.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Michaelsson M, Engle MA. Congenital complete heart block: an international study of the natural history. Cardiovasc Clin. 1972;4:85-101.
2. Gordon PA. Congenital heart block: clinical features and therapeutic approaches. Lupus. 2007;16:642-6.
3. Kannankeril PJ, Fish FA. Disorders of cardiac rhythm and conduction. In: Allen HD, Driscoll DJ, Shaddy RE, Feltes TF, eds. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 7th ed. Philadelphia: Wolters Kluwer/Lippincott Williams & Wilkins; 2008: 293.
4. Buyon JP, Hiebert R, Copel J, Craft J, Friedman D, Katholi M et al. Autoimmune-associated congenital heart block: demographics, mortality, morbidity and recurrence rates obtained from a national neonatal lupus registry. J Am Coll Cardiol. 1998;31(7):1658-66.
5. Costedoat-Chalumeau N, Georgin-Lavialle S, Amoura Z, Piette JC. Anti-SSA/Ro and anti-SSB/La antibody-mediated congenital heartblock. Lupus. 2005;14:660-4.
6. Groves AM, Allan LD, Rosenthal E. Outcome of isolated congenital complete heart block diagnosed in utero. Heart. 1996;75(2):190-4.
7. Groves AM, Allan LD, Rosenthal E. Therapeutic trial of sympathomimetics in three cases of complete heart block in the fetus. Circulation. 1995;92(12):3394-6.
8. Yang CH, Chen JY, Lee SC, Luo SF. Successful preventive treatment of congenital heart block during pregnancy in a woman with systemic lupus erythematous and anti-Sjögren's syndrome A/Ro antibody. J Microbiol Immunol Infect. 2005 Oct;38(5):365-9.
9. Jaeggi ET, Fouron JC, Silverman ED, Ryan G, Smallhorn J, Hornberger LK. Transplacental fetal treatment improves the outcome of prenatally diagnosed complete atrioventricular block without structural heart disease. Circulat. 2004;110:1542-8.
10. Schmidt KG, Ulmer HE, Silverman NH, Kleinman CS, Copel JA. Perinatal outcome of fetal complete atrioventricular block: a multicenter experience. J Am Coll Cardiol. 1991;17:1360-6.
11. Lopes LM, Tavares GM, Damiano AP, Lopes MA, Aiello VD, Schultz R et al. Perinatal outcome of fetalatrioventricular block: One-hundred-sixteen cases from a single institution. Circulat. 2008;118:1268-75.
12. Jaeggi ET, Hornberger LK, Smallhorn JF, Fouron JC. Prenatal diagnosis of complete atrioventricular block associated with structural heart disease: combined experience of two tertiary care centers and review of the literature. Ultrasound Obstet Gynaecol. 2005;26:16-21.
13. Berg C, Geipel A, Kohl T, Breuer J, Germer U, Krapp M et al. Atrioventricular block detected in fetal life: associated anomalies and potential prognostic markers. Ultrasound Obstet Gyanecol. 2005;26:4-15.

DOI: 10.5455/2349-3933.ijam20140508
Cite this article as: Kamath V, Navada M, Dhara, D'Cunha P. Fetal isolated congenital heart block associated with maternal anti-SSA/SSB antibodies. Int J Adv Med 2014;1:37-9.