CASE REPORT

A Case of Prenatal Diagnosis of Congenital Total AV Block on VSD and PDA with Ultrasound

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Abstract

Objective: To report the diagnosis and management of congenital total AV block on VSD and PDA in pregnancy.

Method: Case report

Case: A 37-year-old multiparous woman G4P3A0H2 24- 25 weeks of preterm pregnancy with fetal bradycardia, VSD, PDA with ultrasonography and CTG results was FHR 70 bpm. At 37- 38 weeks of pregnancy, termination of pregnancy was performed by cesarean delivery with preparation for complication of fetal AV block. A male baby was born with weight 2600 gram and APGAR score of 8/9. Immediate echocardiography result was situs solitus, VSD PM L→R shunt, PDA L→R shunt, good left ventricular function, left aortic arch and EF 74%. ECG result was sinus bradycardia, total AV block with junctional escape rhythm. Sternotomy and PPM implantation was performed by cardiothoracic surgeon three hours after the baby was born. Post PPM implantation, ECG results was HR 165 bpm and chest X-rays interpretation was cardiomegaly with plethora. Mother and baby came home in good condition on the 6th day of treatment. On the next baby’s control at 4.5 months obtained a weight of 5.4 kg with the echocardiography results was solitus, VSD PM L→R shunt, VSD muscular multiple 3 pieces L→R shunt, PDA L→R shunt, good right and left ventricular function, and left arch. The child got captopril 2x1.5 mg and planned for a 6-month repeat echocardiography.

Conclusion: Congenital of total AV block on VSD and PDA is confirmed by prenatal diagnosis and preparation for comprehensive multidisciplinary management.

Keywords: congenital total AV block, fetal bradycardia, fetal echocardiography, PPM, ultrasound

BACKGROUND

Fetal bradycardia is defined as a baseline fetal heart rate (FHR) less than 110 beats per minute (bpm) that is present for 10 minutes or longer.1 Bradycardia often being documented during routine ultrasound examination or at the time of routine antenatal visits, requires further investigation to clarify its mechanism.2 Many causes of fetal bradycardia exist (Box 1). Usually these episodes develop abruptly in the setting of a normal FHR baseline and are
followed by a nonstable baseline, decreased or absent variability, and a lack of accelerations and often reflect potential fetal compromise. Fetal bradycardia can also be seen in cases of complete heart block, but, in that setting, the FHR baseline is usually 70–80 bpm with absent variability, and the diagnosis can be confirmed by fetal echocardiography.1

1. Maternal
   a. Hypotension or underperfusion
      i. Position
      ii. Status post-regional anesthesia
      iii. Hemorrhage
      iv. Cardiac dysfunction
   b. Hypoxia
      i. Pulmonary dysfunction

2. Uterine
   a. Tetany (spontaneous or medication induced)
   b. Rupture

3. Placental
   a. Abruption
   b. Infarct

4. Umbilical cord compression
   a. Prolapse
   b. Occult cclusion
   c. Rapid cervical dilation with fetal head descent

5. Fetal
   a. Hemorrhage or anemia
      i. Fetal-to-maternal hemorrhage
      ii. Ruptured fetal vessel (vasa previa)
   b. Hypoxia
   c. Cardiac arrhythmia (heart block)

**Figure 1. Causes of Fetal Bradycardia**

Congenital atrioventricular block (AVB) is a dysfunction of the conducting system and occurs in 1/15,000 to 1/20,000 live births.3 About half of AVBs occur in association with structural heart diseases, such as atrioventricular septal defects, left atrial isomerism and abnormalities of the great arteries.3 The other half is associated with the transplacental transfer of maternal circulatory antibodies against Ro/La antigens.3,4 Anti-SSA/Ro and anti-SSB/La antibodies are closely associated with autoimmune diseases such as systemic lupus erythematosus (SLE), Sjögren syndrome, rheumatic arthritis, and systemic sclerosis, which

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occur frequently in women of child-bearing age. Autoimmune congenital AVB develops only in 1% to 2% of anti-SSA/Ro and/or anti SSB/La positive pregnancies, but its recurrence rate is 12% to 20%. Although uncommon, fetal AVB has significant morbidity and mortality. Complete or total or third-degree AVB appears to be irreversible, but anecdotal reports suggest that treatment of second-degree AVB can restore sinus rhythm.

Congenital AVB is related to significant morbidities such as dilated cardiomyopathy, fetal hydrops and neonatal heart failure. The perinatal mortality rate of congenital AVB varies between studies with a 5.5% to 25% rate of intrauterine fetal demise and a 18% to 40% rate of neonatal mortality. The fundamental treatment of congenital AVB is based on postnatal pacemaker implantation, while there are few options for antenatal management. Fluorinated steroids, such as dexamethasone and betamethasone, have been used to diminish fetal cardiac inflammatory injuries and to increase the fetal heart rate, but some reports do not support their effectiveness.

We report a case of congenital AVB with a structural heart defect (ventricular septal defect and patent ductus arteriosus) which were detected during a midtrimester screening ultrasound who treated by post natal permanent pacemaker implantation.

CASE

A 37 year-old multiparous woman was referred to our institution at 37-38 weeks of pregnancy for fetal bradycardia detected by routine midtrimester screening ultrasound. History ultrasound at private clinic at 24-25 weeks of pregnancy and known as fetal bradicardia with VSD and PDA. There was no evidence of fetal hydrops or other fetal structural abnormalities. The patient had no symptoms of autoimmune diseases, such as skin rash, joint pain, photosensitivity, alopecia, and oral ulcers. A work up for maternal TORCH infection and autoimmune disease screening. The result is positive CMV infection. Before delivery, we checked the fetal heart with ultrasonography weekly at the outpatient clinic. The ultrasonographic findings showed no significant changes.
An elective lower segment cesarean section was performed at 37-38 weeks of pregnancy with preparation for complication of fetal AV block. A male baby born was born with weight 2.6 kg at birth, and the Apgar scores at 1 and 5 minutes were 8 and 9, respectively. The baby’s heart rate was measured under 100 beats per minute and the lower rate 40 to 50 beats per minute on an monitor. ECG results was ECG result was sinus bradycardia 75 bpm, total AV block with junctional escape rhythm.

Immediate echocardiography was performed with the result solitus site, concordance A-V and V-A connection, all PV to LA, balance ventricle with good LV and RV function, IAS bulging to LA, VSD perimembranous diameter 3 mm L-R shunt, PDA 3-4 mm L-R shunt, left arch and the cardiac team decided to perform insertion permanent pacemaker (PPM) with surgical approach. The primary choice was dual chamber pacemaker. But for some reason, cardiologist decide to use single chamber pacemaker VVIR mode.
Sternotomy and PPM implantation was performed by cardiothoracic surgeon three hours after the baby was born. A single chamber pacemaker VVIR mode with unipolar lead was inserted in epicardial RV, lower rate 130 bpm upper rate 170 bpm amplitude 3.5 Volt, pulse width 0.4 ms, sensitivity 2.8 mV, Threshold 1 V, R wave 15.6, lead impedance 481 ohms, battery longevity 3-5 years. Post pacemaker implantation, ECG results was HR 165 bpm. Chest x-ray was performed and showed a cardiomegaly with CTR 66% with plethora.

Mother and baby came home in good condition on the 6th day of treatment. On the next’s baby’s control at 4.5 months obtained a weight of 5.4 kg with the echocardiography results was solitary, VSD PM L-R shunt, VSD muscular multiple 3 pieces L-R shunt, PDA L-R shunt, good right and left ventricular function and left arch. The child got captopril 2x1.5 mg and planned for 6-month repeat echocardiography.

DISCUSSION

Elucidating the underlying electrophysiological mechanism leading to fetal bradycardia is of the utmost importance, as management strategies vary and depend on achieving a correct diagnosis. The most frequently used ultrasound techniques in clinical practice are M-
mode echocardiography and pulsed wave Doppler. Other diagnostic modalities, such as tissue Doppler imaging, fetal electromagnetocardiography, may also help but are less commonly used. Bradycardia can be regular or irregular. Regularity can be ascertained by simply listening to the fetal heart or by visual inspection of M-mode or Doppler recordings, which should show equal time intervals between ventricular contractions (regular V-V interval).

Congenital AVB is a very rare congenital anomaly, but carries significant fetal or neonatal mortality and morbidities. Congenital AVB can be diagnosed in utero, at birth or within the neonatal period, but it commonly develops during the 18 to 24 weeks of pregnancy. Because of the rapid progression of the disease from 1st or incomplete block to 2nd and 3rd block, most of the cases are not detected until they develop complete or total AVB. Congenital atrioventricular block is a term used to describe complete atrioventricular block that occurs spontaneously and is diagnosed in a fetus, at birth, or within the first month of life. There are three major etiologies of congenital atrioventricular block structural heart defects in the setting of congenital heart disease, antibody-mediated congenital heart block and idiopathic congenital atrioventricular block.

The first etiology of congenital AVB is manifest with various forms of congenital heart disease. These congenital heart defects are associated with developmental abnormalities of the conduction tissue. Displacement of the compact AV node in a posterior direction is often seen in patients with endocardial cushion defects, for example. Additionally, the AV node in a patient with endocardial cushion defects may be less robust resulting in a more rapid deterioration in function over time, but also a higher predilection for surgical or mechanically induced AV block. Patients with congenitally corrected transposition or L-looped TGA also have a displaced and less robust AV node.

The second etiology is the most common etiology (60-90% of cases) of congenital AVB is associated with placental transference of maternal Ro/La autoantibodies, in which case the disease is known as auto-antibody mediated, Ro/La related or autoimmune-mediated congenital AVB. The most common maternal diagnoses in babies born with autoimmune mediated congenital AVB include Sjogren’s syndrome (SS) or systemic lupus erythematosus (SLE). These Ro/La autoantibodies enter the fetal circulation beginning in the mid-second trimester, and can result in damage to the conduction tissues during fetal development including inflammation, calcification and fibrosis, leading to conduction block at the level of AV node in an otherwise structurally normal heart. The first sign of our case was fetal bradycardia with a ventricular rate of 70 bpm and complete block at 24-25 weeks of pregnancy. This is the typical presentation of congenital AVB. Some predictive factors of poor outcomes are suggested, including the presence of a congenital structural heart defect or fetal hydrops, and early gestational age at diagnosis. The clinical phenotype of congenital AVB
is variable and dependent on an assortment of factors including the underlying etiology, age of the patient at the time of presentation, the patient’s escape rate, and the ventricular function. Those patients with autoimmune congenital AVB can be identified in utero before 30 weeks gestation and tend to present earlier than patients with congenital AVB secondary to structural or idiopathic etiologies.\textsuperscript{10}

In addition, patients with autoimmune associated congenital AVB can present with other cardiac sequelae. In newborns, bradycardia with a documented heart rate less than 100 beats per minute is usually the primary finding. Other clinical findings may include symptoms and signs of congestive heart failure. In comparison to older children and adults, first or second degree AV block found in infants should be monitored for progression to CHB. In older children, the primary finding is a bradycardia with or without symptoms including exercise intolerance, presyncope or syncope.\textsuperscript{13}

For the fetal diagnosis of congenital AV block, the echocardiography has been determined as the gold standard, however, all M-mode and pulsed wave Doppler echocardiographic techniques that are based on the relationship between the atrial and ventricular mechanical events, can be well applied, besides clinical application of fetal electrocardiography and fetal magnetocardiography are promising noninvasive and highly precise tools.\textsuperscript{12} Elucidating the underlying electrophysiological mechanism leading to fetal bradycardia is of the utmost importance, as management strategies vary and depend on achieving a correct diagnosis. Other diagnostic modalities, such as tissue Doppler imaging, fetal electromagnetocardiography, may also help but are less commonly used.\textsuperscript{2} Bradycardia can be regular or irregular. Regularity can be ascertained by simply listening to the fetal heart or by visual inspection of M-mode or Doppler recordings, which should show equal time intervals between ventricular contractions (regular V-V interval).\textsuperscript{2}

Referent to the treatment, dexamethasone is currently used, a study in which was included maternal treatment with dexamethasone was associated with normalized AV conduction in fetuses with first-degree AV Block, however its administration is also associated with potential side effects for both mother and fetus, especially potential negative effects in osteogenesis in the embryo developing.\textsuperscript{14,15,16}
A permanent pacemaker placement is needed in most children with congenital heart block. The medical care of congenital heart block is currently focused on identifying the optimal timing of pacemaker therapy to ensure a positive outcome.\textsuperscript{17,18} Pacemaker implantation may improve long term survival as well as prevent syncopal events among those asymptomatic patients with congenital AVB. In those patients diagnosed in utero, management options are limited. Fortunately, slow escape rhythms leading to fetal distress and hydrops is rare, with the majority of patients having an adequate rate to support their hemodynamics through term gestation. Pharmacological maneuvers to alter the AV nodal conduction or enhance the fetal heart rate have limited efficacy.\textsuperscript{15} In this case, the primary choice was dual chamber pacemaker. But for some reason, cardiologist decide to use single chamber pacemaker VVIR mode.\textsuperscript{19,20}
CONCLUSION

Fetal bradycardia can be diagnosed in the prenatal period. The most frequently used ultrasound techniques in clinical practice are M-mode echocardiography and pulsed wave Doppler. Fetal bradycardia of primary cardiac origin can be a benign finding if it is due to blocked ectopic beats. These have no hemodynamic consequence, are well-tolerated by the fetus and have no long-term consequences. Conversely, fetal bradycardia that is linked to pathological AV block is often an autoimmune process. It can affect myocardial function as well as the conduction tissue. A permanent pacemaker placement is needed in most children with congenital AV block.

REFERENCES


