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CASE REPORT

Prenatal diagnosis of complete atrioventricular block secondary to maternal lupus: a case report

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Abstract

This report describes the case of a newborn with congenital complete atrioventricular (AV) block diagnosed antenatally belonging to a mother diagnosed with lupus erythematosus during pregnancy based on an echocardiogram ordered to investigate a finding of slow fetal heart rate. Pregnancy was terminated at 36 weeks with a cesarean section indicated due to preeclampsia. Neonatal resuscitation protocols and vasoactive drugs did not increase the newborn's heart rate to above 100 beats per minute. The newborn had a pacemaker implanted 20 days after birth and recovered well. He was discharged without other measures to maintain heart rate. The disease was attributed to previously undiagnosed maternal systemic lupus erythematosus (SLE), which was investigated once the fetus was diagnosed with complete AV block. The success of the procedure derived from an accessible prenatal care program and an efficient public healthcare system.

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INTRODUCTION

Differently from adults diagnosed with systemic lupus erythematosus, electrical cardiac abnormalities such as atrioventricular (AV) blocks are known immunological complications in newborns of mothers with anti-Ro and anti-La autoantibodies, often seen in women with systemic lupus erythematosus (SLE) or Sjögren's syndrome[1]. Sixty to ninety percent of the cases of AV block are attributed to these conditions. In the general population, the incidence of congenital complete AV block is about 1 per 15,000 to 20,000 live births[2]. Approximately 2% of the children exposed to anti-RO and anti--LA antibodies develop complete AV block[3].

Two factors influence the survival of fetuses affected by neonatal lupus: early detection and treatment of AV blocks. According to the Brazilian Guidelines for Fetal Echocardiography, the test of choice for this evaluation is fetal Doppler echocardiography[4].

CASE REPORT

A 31-year-old primigravida started prenatal care at a basic healthcare clinic in the first trimester. She was on fluoxetine for anxiety and clonazepam for seizures, and had no other complaints or diagnosed conditions. During prenatal care, ultrasound examination performed on the 21st week of gestation detected a fetal heart rate of 52 bpm. A fetal echocardiogram was ordered promptly and performed on the 26th week of pregnancy, at which time the fetus was diagnosed with complete AV block. The patient was referred to a prenatal service for high-risk pregnancies and the rheumatology service of a university hospital.

The rheumatologist sought potential immune cases for the complete AV block of the newborn. A detailed interview was performed, in which the mother's complaints of bilateral joint pain in the knees, sensation of heaviness in the ankles, asthenia, erythematous itchy painful lesions throughout her body, and bilateral hyperemia in the malar area after exposure to sun light were analyzed. She had these symptoms more than a year prior to becoming pregnant, but they were neglected by her and her doctors. ANA and anti-Ro antibody tests were ordered and came back with positive, high titers, confirming the diagnosis of SLE associated with Sjögren's syndrome after salivary gland scintigraphy and ophthalmologic evaluation. The Rheumatology team of the Brasília University Hospital prescribed hydroxychloroquine during the final periods of gestation, resulting in partial remission of symptoms. She was followed-up as a high-risk patient with weekly echocardiograms. In the test performed four days before delivery, findings consistent with atrioventricular dissociation were observed without significant signs of heart failure.

Since she was diagnosed with preeclampsia, delivery was scheduled for 36 weeks and 3 days of gestation (36 weeks and 2 days according to the Capurro method). Her male preterm newborn was deemed as having low birth weight (2460g) and being adequate for gestational age. He had central cyanosis at birth, irregular crying, weak muscle tone, and a heart rate below 80 bpm. He was positioned under a radiant heat source, dried, and stimulated without improvement. His upper airways were cleaned with the aid of suction, but he remained hypotonic and with central cyanosis. In view of the possibility of his heart rate remaining at levels below 100 bpm and the lack of response to initial resuscitation steps, the newborn was intubated. His tome improved, along with the color of his skin, he began to respond well to stimuli, and his heart rate remained at 50-60 bpm. His 1- and 5-minute APGAR scores were 5 and 7, respectively.

He was taken to the neonatal intensive care unit (NICU) and a central line was placed to deliver a continuous infusion of adrenaline (0.2mcg/Kg/min) and fentanyl (1 mcg/Kg/h). He was kept on a ventilator on a lower setting. Other signs of neonatal lupus were not seen on physical examination: his fontanel measured 2x2 cm; lung auscultation was unaltered; he had no masses or visceromegaly on examination of the abdomen; the patient's skin was intact and without lesions; cardiac auscultation showed an irregular rhythm and bradycardia; perfusion was borderline with capillary refill time of 3 seconds; his pulse was weak. The newborn was on continuous adrenaline at the time of admission. An electrocardiogram showed atrial sinus rhythm with atrioventricular dissociation, a ventricular rate of about 75 bpm, and a normal QRS complex (Figure 1). An echocardiogram taken two days after birth showed a patent foramen ovale and mild aortic insufficiency, without signs of heart failure.

Twenty days after birth, the patient underwent an open-heart surgery to have a pacemaker implanted. Until then, he was kept on continuous adrenaline with a target heart rate above 50 bpm. He improved satisfactorily after surgery, with an increase in heart rate to between 115-119 bpm. Tissue perfusion improved without the use of vasoactive drugs. He was discharged on exclusive breastfeeding five days after surgery with scheduled outpatient follow-up visits.

DISCUSSION

The case described in this report draws attention to the diagnosis of complete AV block in a fetus performed that of the mother. Congenital AV block is the most common and severe clinical manifestation of neonatal lupus, affecting about 2% of children with the condition, with an estimated recurrence rate of 20% in subsequent pregnancies[3]. The mothers of these children are mostly asymptomatic (about 50%); some have SLE or Sjögren's syndrome[3]. The patient's mother was believed to be healthy. However, a closer review of her medical history indicates the existence of overlooked symptoms consistent with SLE, a condition she was diagnosed with after her son's electrical disorder was found. The manifestations of neonatal lupus can be diverse and involve multiple systems, as reported by Freire et al.[5]

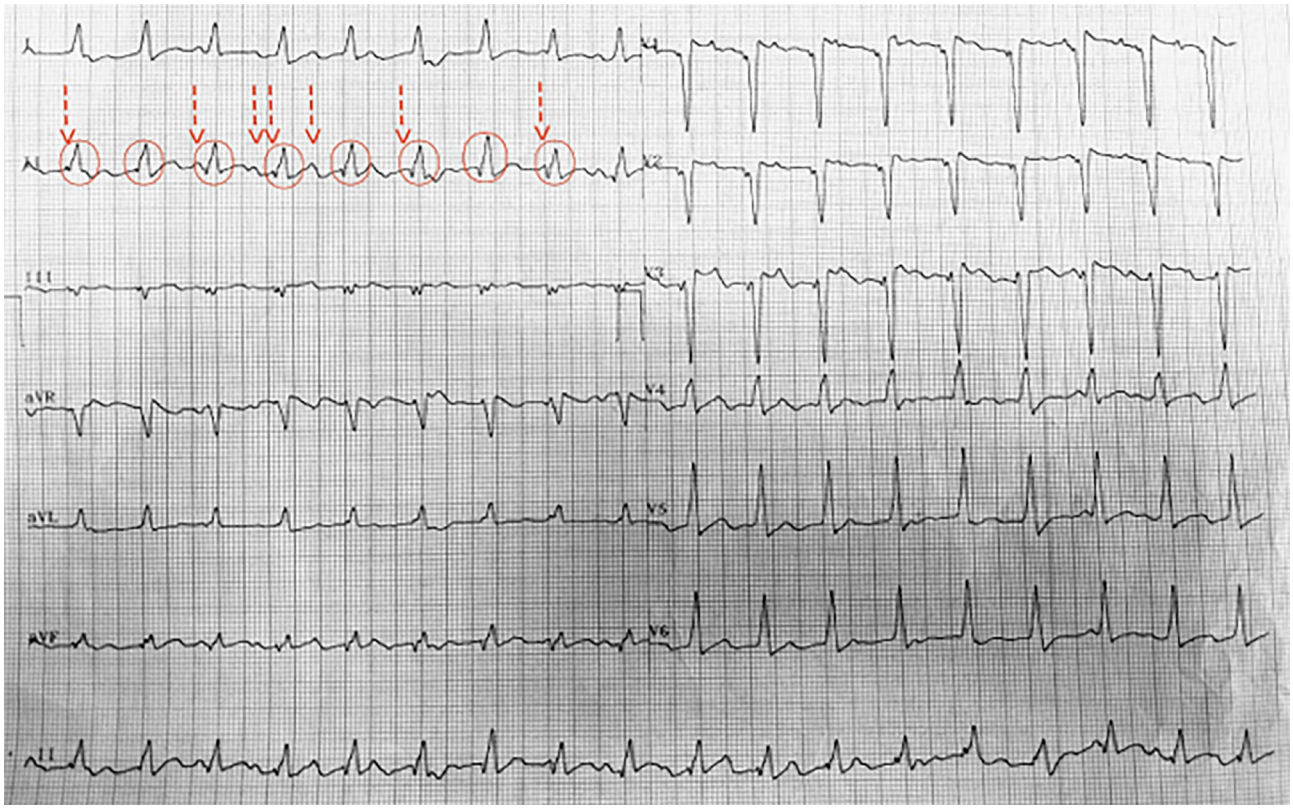


Figure 1. ECG before intervention ECG image showing complete atrioventricular block: atrial sinus rhythm with atrioventricular dissociation, ventricular rate of about 75 bpm, normal QRS complex. The arrows point to the P waves that are dissociated from the QRS complexes shown in the circles.

The pathophysiology of neonatal lupus includes several maternal, fetal, and environmental components[6]. When present in the mother, autoantibodies such as anti-RO and anti-LA can trigger an inflammatory response in the fetal myocardium, which may culminate with cardiac conduction defects, myocarditis, endocardial fibroelastosis, and heart failure[7]. Conduction system cells are preferentially involved, especially in the atrioventricular node, the sinoatrial node, and the bundle of His, which explains the appearance of fetal heart conduction alterations. Genetic predisposition and susceptibility of the fetus to maternal autoantibodies must also be considered[8]. Analysis of heart tissue of fetuses that died of AV block found cardiomyocyte apoptosis and macrophage clusters in fibrotic areas with the simultaneous presence of IgG and tumor necrosis factor, extensive deposition of collagen in the conduction system, lymphocyte infiltrate, and calcification[7].

Other studies have also described an association between anti-Ro antibodies and complete AV block outcomes. Brucato et al.[3] reported two cases of complete AV block out of 122 newborns from 100 women with anti-Ro antibodies followed during pregnancy. Interestingly, the mother of children with complete AV block had Sjögren's syndrome and mixed connective tissue disease. The author recognizes that most pregnant women with fetuses with complete AV block are asymptomatic or oligosymptomatic, as seen in the case herein presented.

In a retrospective study, Buyon et al.[6] included 57 women with children diagnosed with congenital heart block, and found anti-Ro antibodies in the ELISA tests of 100% of the women, showing the important association between this antibody and congenital complete AV block. The antibody titers seen in individuals with positive test results seem to be involved in the neonatal development of AV block. The Brazilian guidelines for fetal echocardiography recommend that previously healthy mothers pregnant with fetuses with congenital complete AV block be tested for these antibodies[4]. In our case, the mother had Anti-Ro > 240 U/ml (RV > 10.1 U/ml) at 25 weeks of gestation, and the fetal echocardiogram showed a structurally normal heart without signs of heart failure, but asynchrony of atrial and ventricular beats with a ventricular rate of 59 bpm.

According to the guideline cited above, after a diagnosis of complete AV block and a positive maternal anti-RO antibody test, in case of a sinus rhythm, the AV interval must be followed weekly from the 18th to the 26th week of gestation with pulse-Doppler ultrasound. Additionally, myocardial function must be monitored every four weeks[4]. We could not follow the AV interval in our patient as recommended, since there was a delay in the diagnosis of the disorder due to the fact that the patient was oligosymptomatic.

The following three therapeutic pillars are currently applied for the treatment of immune-mediated congenital AV

block: immunoglobulin, steroid therapy, and pacing. The severity of complete AV block calls for the institution of prophylactic therapies. According to the Brazilian Fetal Echocardiography Guidelines[4], dexamethasone can be used in cases in which the AV interval exceeds 150 milliseconds, with doses varying from 4 to 8 mg; it can also be used in cases in which complete AV block is installed positive test results for antibodies and immunoglobulin are present. Benefits include inflammation reduction and potential reversion or stabilization of fetal hydrops, endocardial fibroelastosis, and 1st and 2nd degree blocks[4]. The association of the two medications may increase the survival of fetuses with endocardial involvement or systolic dysfunction. There is no recommendation supporting the prophylactic use of these medications. Salbutamol, terbutaline, or isoprenaline can be used to increase the heart rate of fetuses with the condition. However, there is no evidence of increased survival, although it may prolong gestation. Finally, intrauterine pacemaker implantation lacks evidence and has been attempted in fetuses with severe disease. Use of steroids in pregnancy must be well indicated, given the large number of possible side effects such as intrauterine restricted growth, oligohydramnios, adrenal suppression, learning disorders, reduced brain growth, and diabetes[9].

Finally, according to the Brazilian Guidelines on Implantable Electronic Heart Devices from the Artificial Heart Stimulation Department of the Brazilian Society of Cardiovascular Surgery[10], pacemaker implantation in individuals with congenital complete AV block is indicated when the patient is symptomatic or has a heart rate below 50 bpm at rest or below 70 bpm associated with structural heart disease. Our patient was unable to sustain sufficient cardiac performance and maintain hemodynamic stability while in the delivery room, which prompted the initiation of mechanical ventilation and administration of vasoactive drugs to avoid cardiovascular collapse. Although structural alterations were absent and his heart rate was above 50 bpm, since he had symptoms, the implantation of a pacemaker was a good decision.

The in-utero diagnosis of congenital complete AV block was imperative for the adequate preparation of the healthcare team involved in the delivery of a newborn with cardiac arrhythmia, and allowed prompt evidence-based intervention, the initiation of emergency therapy administration, and the scheduling of pacemaker implantation surgery.

CONCLUSION

This case stresses the importance of prenatal care in the early detection and treatment of disorders to mitigate maternal, fetal, and neonatal mortality and morbidity. Although uncommon, cases of neonatal AV block due to neonatal lupus are relatively easy to diagnose with adequate prenatal follow-up. Timely diagnosis allows for adequate planning of the interventions needed to preserve the lives of mothers and children. Building strong physician-patient relationships may reduce the morbidity associated with poor early detection of diseases due to signs and symptoms missed during medical visits.

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