

# Fetal Arrhythmia; Maternal Systemic Lupus Erythematosus

Pınar TOKDEMİR ÇALIŞ<sup>1</sup>, Süheyla ÖZKUTLU<sup>2</sup>, M. Sinan BEKSAÇ<sup>1</sup>

Ankara, Turkey

Congenital heart block may develop in the fetus during pregnancy in anti-Ro/La antibodies positive women, leading to heart failure of fetus in utero and even mortality in the first 3 years of life. In this study, we report a fetal arrhythmia case who manifested in 21 weeks of gestational age. Diagnosis of Systemic Lupus Erythematosus (SLE) was confirmed with the positive blood samples for antinuclear antibodies, anti-Ro/SSA and anti-La/SSB and ultrasonography. There was no hidrops in the fetus when arrhythmia was detected. After steroid therapy, patient's symptoms concerning SLE regressed. Following the delivery of baby, permanent pacemaker had been applied and currently, the baby has no cardiac problem. Therefore, it could be concluded that, SLE can be asymptomatic till pregnancy and the control of the disease is essential for the delivery of a healthy baby.

**Key words:** Congenital heart block, Fetal arrhythmia, Systemic lupus erythematosus (SLE), Pregnancy, Anti-Ro/la antibodies

Gynecol Obstet Reprod Med 2011;17:177-179

## Introduction

Congenital heart block affects 2% of all pregnant women who have anti-Ro/La antibodies positivity, leading to heart failure in utero, and has a 20% mortality rate in the first 3 years of life.<sup>1</sup> These autoantibodies are mostly found to be positive in patients with SLE and Sjogren Syndrome. The aim of this case report is to analyze the management of newly diagnosed fetal arrhythmia in pregnant patients.

## Case Report

A 21 year-old pregnant woman presented to our hospital at 34 weeks of gestational age, after the diagnosis of fetal arrhythmia initially manifested at 21 weeks of gestational age. Her gravida was 2. She had one intrauterine exitus at the first trimester, in her previous pregnancy. Beginning with her first pregnancy, she had intermittent pain in her joints. In this pregnancy, first admission to the obstetrician was at the 8<sup>th</sup> week of pregnancy. There had been no detected developmental problem of the fetus during the routine controls until 21<sup>st</sup> week of

pregnancy. Then she was diagnosed with fetal arrhythmia during routine ultrasonography. After a follow up period she was referred to our obstetrics clinic at the 34 weeks of gestational age, because of fetal arrhythmia. In addition to the fetal arrhythmia she complained of pain in her bilateral hands, proximal phalanges, and knee joint. She also had perihepatic ascites, edema of all parenchymal organs and skin. Some laboratory tests, especially autoantibodies, were performed. Mother showed high titers of anti nuclear antibody - ANA(1/1000), ENA SsA (1039IU/mL), ENA SsB (1181IU/mL). Her complement 3 and 4 levels were detected in normal ranges. Her vitamin B12 level was measured to be low (92pg/mL). In addition MTHFR 677 heterozigot gene mutation was detected. For detailed analysis of fetal arrhythmia Doppler ultrasound was performed besides routine obstetric USG revealing third degree fetal atrio ventricular (AV) block.

The lab results confirmed the diagnosis of SLE and with respect to the fetal third degree heart block steroid therapy with oral dexamethasone and methylprednisolone 4 mg.each daily (in two equal doses) initiated. After 10 days of treatment with oral steroids, the pain in her knees, and finger joints were decreased, and perihepatic ascites resolved. The pregnancy continued uneventfully and a 2500gr healthy female infant was delivered at 37 weeks and 5 days of gestation with elective cesarean section. Implantation of a permanent rate modulated single chamber ventricular endocardial pacemaker was performed a few days after the delivery. A regular sinus rhythm at a rate of 120-130 bpm with a normal PR interval was noted. At the infant's last follow up visit at age 6 months revealed no cardiac problem.

<sup>1</sup>Departments of Obstetrics and Gynecology and <sup>2</sup>Pediatrics Hacettepe University Medical Faculty, Ankara

Address of Correspondence: Prof.Dr. M. Sinan Beksaç  
Hacettepe Üniversitesi Tıp Fakültesi  
Hastanesi Kadın Hastalıkları ve Doğum  
Ana Bilim Dalı Ulus, Ankara  
beksaç@tr.net

Submitted for Publication: 26. 03. 2011

Accepted for Publication: 06. 07. 2011

## Discussion

There are three types of fetal arrhythmias. The most common form is irregular heartbeat, mainly caused by ectopic beats. Other two types of arrhythmias are fetal bradycardia and tachycardia.<sup>2</sup> Fetal bradycardia is diagnosed with the fetal heart rate slower than 100bpm. which is mainly due to AV block.<sup>2</sup>

Congenital complete heart block (CHB) was first described in 1901 by Morquio, who has also noted the familiar occurrence and association with Stokes-Adams attacks and death.<sup>3</sup> Neonatal lupus is a passively transferred autoimmune disease that occurs in some babies born to mothers with anti-Ro/SSA and/or anti-La/SSB positive. These antibodies commonly found in people with Sjogren's disease and SLE. They could be also found in patients with rheumatoid arthritis, progressive systemic sclerosis, cutaneous vasculitides.<sup>4</sup> Neonatal lupus, due to maternal antibodies, especially SS-A that cross the placenta, is reported to be responsible for 60 to 90 percent of cases of CHB overall.<sup>5</sup> Almost all cases are presenting in utero or neonatal period, only %5 of the cases have been reported to occur later.<sup>5</sup>

Anti-Ro/SSA and anti-La/SSB antibodies bind to fetal cardiac tissue, causing an autoimmune injury in the atrioventricular (AV) node and its surrounding tissue. These antibodies are both abundant in fetal heart tissue between 18 and 24 weeks.<sup>6</sup> By this mechanism, apoptosis is induced at the fetal cardiomyocytes and fibrosis occurs. These antibodies also inhibit calcium channel activation by this way and inhibit the conduction in the AV node.<sup>7</sup> As a result, congenital heart block may present with fetal bradycardia between 18 and 28 weeks of gestation.<sup>5,8</sup> Detailed analysis of the type of arrhythmia in utero is possible using M-mode and Doppler echocardiography.<sup>2</sup>

For our patient, anti-Ro/SSA and anti-La/SSB antibodies were determined as both positive. There were typical SLE symptoms in her previous pregnancy but she was not diagnosed. Also the fetal bradycardia presented at 21 gestational week is also a common presentation according to the previous reports.<sup>2</sup>

Management of congenital heart block in utero and in the perinatal period can include steroid therapy if associated with anti-Ro/SSA and anti-La/SSB antibodies and pace maker or isoproterenol administration immediately at the postpartum period.<sup>9</sup> In some cases, AV block can result with fetal hidrops. Some clinical studies reported that,<sup>9</sup> steroids have no effect on hidropic fetus.<sup>10</sup> On the other hand, in our case fetal hidrops was not observed. For these reasons, oral dexamethasone and methylprednisolone therapy was initiated.

In conclusion, SLE can be asymptomatic till pregnancy

and the control of the disease is essential for the delivery of healthy baby. According to study of Wei et. Al<sup>11</sup> twenty-four (44.44%) pregnant woman with anti-Ro/SSA and/or anti-La/SSB antibodies were asymptomatic and antibody status is first indicated when offspring shows symptoms of Neonatal Lupus Erythematosus. It is considered that it might be a better approach that all pregnant women are suggested for screening anti-Ro/SSA and anti-La/SSB antibodies when they are administered to the hospital first time during their pregnancies.

## Fetal Aritmi; Maternal Sistemik Lupus Eritematosus

Anti-Ro/La antikorlarına sahip gebe bir kadının bebeğinde konjenital kalp bloğu oluşabilir ve bu blok anne karnında bebeğin kalp yetmezliğine girmesine ve hatta bebeğin yaşamının ilk üç yılında mortaliteye sebep olabilir. Bu çalışmada, 21. gebelik haftasında tanı almış bir fetal aritmi vakası yayınlanmıştır. Sistemik Lupus Eritematosus (SLE) tanısı kan testlerinde anti-nükleer antikör, anti-Ro/SSA ve anti-La/SSB antikörlerinin pozitif gelmesi ve ultrasonografi ile koyulmuştur. Vakada aritmi saptandığında fetusun hidropsu yoktu. Hastaya verilen steroid tedavisinden sonra, hastanın SLE ile ilgili semptomları geriledi. Doğumu takiben, bebeğe kalıcı kalp pili takıldı ve takiplerinde bebeğin kardiyak herhangi bir problemi yoktur. Bundan dolayı, şu sonuca varılabilir ki, SLE gebeliğe kadar asemptomatik kalabilir ve bu hastalığın kontrolü ve tedavisi sağlıklı bir bebek dünyaya getirmek için çok önemli ve şarttır.

**Anahtar Kelimeler:** Konjenital kalp bloğu, Fetal aritmi, Sistemik lupus eritematosus (SLE), Gebelik, Anti-Ro/La antikörleri

## References

1. David A. Atallah I. Yates R. Congenital fetal heart block: a potential therapeutic role for intravenous immunoglobulin. Institute for Women's Health, University College London Hospitals, London, United Kingdom.
2. Maeno Y, Hirose A, Fetal arrhythmia: Prenatal diagnosis and perinatal management. J. Obstet. Gynaecol. Res. 2009;35:623-9
3. White, P, Eustis, R. Congenital heart block. Am J Dis Child 1921;22:299
4. Cavazzana I. Franceschini F. Belfiore N. et al. Undifferentiated connective tissue disease with antibodies to R/SSA; Clinical features and follow-up of 148 patients. Clin Exp Rheumatol 2001;19:403.
5. Jaeggi,ET, Hamilton, RM, Silverman, ED, et al. Outcome of children with fetal, neonatal or childhood diagnosis of isolated congenital atrioventricular block. A single institution's experience of 30 years. Jam Coll Cardiol 2002; 39:130.
6. Alexander, E, Buyon, JP, Provost, TT, et al. Anti Ro/SS-A

- antibodies in the pathophysiology of congenital heart block in neonatal lupus syndrome. An experimental model. *Arthritis Rheum* 1992;35:176.
7. Miranda-Caru, ME, Askanase, AD, Clancy, RM, et al. Anti-SSA/Ro and anti-SSB/La autoantibodies bind the surface of apoptotic fetal cardiocytes and promote secretion of TNF-alpha by macrophages. *J Immunol* 2000;165:5345.
  8. Buyon JP, Hiebert R, Copel J, et al. Autoimmune-associated congenital heart block: demographics, mortality, morbidity and recurrence rates obtained from a national neonatal lupus registry. *Am Coll Cardiol* 1998; 31:1658.
  9. Michaelsson M, Engle MA. Congenital complete heart block: An international study of the natural history. *Cardiovasc Clin* 1972;4:85.
  10. Jaeggi E, Fouron J, Silverman E. Transplacental fetal treatment improves the outcome of prenatally diagnosed complete atrioventricular block without structural heart disease. *Circulation*. 2004;21;110:1542-8.
  11. Wei S., Yuan TM., Chen LH, Yu HM. Neonatal lupus erythematosus: three case reports and review of the Chinese literature. *Clin Pediatr (Phila)* 2010;49:627-34.