

## Permanent pacemaker implantation in complete congenital fetal atrioventricular (AV) block: a case report

*Implantación de marcapasos permanente en bloqueo atrioventricular (AV) fetal congénito completo: reporte de caso*

Lydia Enith Nava-Rivera,\* Rodrigo Lozoya-Martínez,\*\*  
Daniel Chi-Argüelles,\*\*\* Javier Morán-Martínez\*\*\*\*

**Key words:**

Pacemaker,  
congenital heart  
block, anti-Ro/SSA,  
anti-Smith, neonatal  
lupus.

**Palabras clave:**

Marcapasos  
permanente, bloqueo  
cardíaco congénito,  
anti-Ro/SSA,  
anti-Smith, lupus  
neonatal.

**ABSTRACT**

One of the most common pathologies attributed to lupus neonatal refers to atrioventricular (AV) congenital block, which diagnosis can be made between 16 and 30 weeks of gestation due to persistent fetal bradycardia. The development of this disease is mostly related to maternal anti-Ro/SSA and anti-Smith autoantibodies. Currently, there are a number of alternatives for prenatal and postnatal treatment, with some controversy about their viability. The placement of a permanent pacemaker is presented as one of the most appropriate procedures currently, even with the risks awarded. This case report describes the placement of a permanent pacemaker to a two-month-old newborn with high maternal contents of anti-Ro/SSA and anti-Smith nuclear autoantibodies, with a favorable outcome.

**RESUMEN**

Una de las patologías más comunes atribuidas al lupus neonatal se refiere al bloqueo congénito atrioventricular (AV), diagnóstico que se puede realizar entre 16 y 30 semanas de gestación debido a bradicardia fetal persistente. El desarrollo de esta enfermedad se relaciona principalmente con los anticuerpos anti-Ro/SSA materno y anti-Smith. Actualmente, existen varias alternativas para el tratamiento prenatal y postnatal, con cierta controversia sobre su viabilidad. La colocación de un marcapasos permanente se presenta como uno de los procedimientos más adecuados actualmente, incluso con los riesgos adjudicados. Este relato de caso describe la colocación de un marcapasos permanente en un recién nacido de dos meses con alto contenido materno de autoanticuerpos anti-Ro/SSA y anti-Smith, con un resultado favorable.

**INTRODUCTION**

The complete AV block that produces a dissociation of atrial and ventricular contractions,<sup>1</sup> is a heart pathology associated with the trans-placental transmission of maternal antinuclear antibodies (ANA's) being a severe manifestation of neonatal lupus.<sup>2</sup> Besides, the cardiac lesions most frequently associated to this pathology are the isomerism of the left atrial appendages (LAI) and discordant atrioventricular connections, affecting about 60% of the diagnosed cases. During pregnancy women detected with ANA's particularly anti Ro/SSA, La/SSB and anti-Smith antibodies have been linked to fetal heart damage.<sup>3-6</sup> Seropositive Ro/SSA or La/SSB woman have

a 2-3% of probability of getting a child that develops congenital AV block or abnormalities of the myocardium<sup>7</sup> and a 17-20% of risk in case of having a previous child with the condition, this situation have its relevance in the prognosis for each case. Complete AV block involves an immune mediated inflammatory process which results in a damage of the conduction tissue and myocardium during the heart developing of the product.<sup>8</sup> For the diagnosis of this pathology the use of ultrasound technology during prenatal care allows an early finding between 18 and 24 weeks of gestation, propitiating a better prognosis and for defining an adequate therapy.<sup>8,9</sup> The principal treatment for a congenital heart block is aimed to a prophylactic sense that

\* Postgraduate Student from the Faculty of Medicine.

\*\* Medical Intern in the Department of Biology and Ultrastructure, Biomedical Research Center. Faculty of Medicine.

\*\*\* Pediatric Cardiologist from the Department of Neonatology, University Hospital of the Faculty of Medicine.

\*\*\*\* Research Professor in the Department of Biology and Ultrastructure,

Biomedical Research  
Center. Faculty of  
Medicine.

Autonomus University  
of Coahuila, Campus  
Torreón.

Received:

07/07/2017

Accepted:

29/08/2017

includes maternal administration of steroids, plasmapheresis, sympathomimetic, and *in utero* cardiac pacing during an adequate gestational age,<sup>10</sup> added to this, a prenatal progression of the cardiac anomaly is important in case that a perinatal management could be necessary, since a pacemaker is generally needed in most children with congenital heart block.<sup>11</sup> Due to this situation, the medical treatment for this pathology is focused on identifying the optimal timing for the placement of a pacemaker in order to ensure a positive outcome.<sup>12</sup>

### CASE REPORT

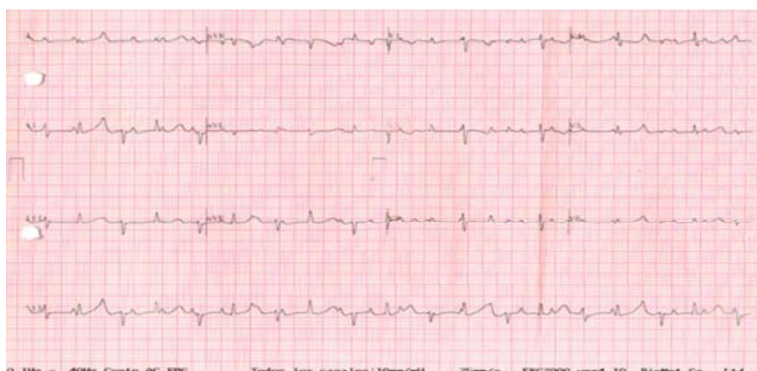
The case refers to a 22 years old patient (G3P2C0A0) who was admitted to the department of gynecology and obstetrics of this hospital with 31.3 weeks period of gestation (POG) and a probable lupus nephropathy, without previous history of smoking or alcoholism, prenatal control started at the second trimester, without complications during pregnancy, with a fetal heart rate between 51 and 105 bpm.

Results of the hematological parameters of the patient resulted in a reduction in erythrocyte mass, hemoglobin and hematocrit levels (2.89  $10^6$ /mL, 8.10 g/dL and 26.5% respectively). The liver function panel resulted in 5 U/L of TGP, high levels of CGT 102 U/L and LDH 1,288 U/L; a renal biopsy was performed showing morphologic changes compatible with diffuse lupus nephritis class IV-S (A). In the

analysis of ANA's the results were positive for anti-Sm and anti-Ro/SSA (166.3 U/mL, 160.64 U/mL respectively). Obstetric echocardiogram showed a dissociation in the heart rhythm with apparent atrioventricular block, with an atrial frequency in the range of 130-140 bpm and at ventricular level with a bradycardia of 58 bpm, without a septal defect or cardiomegaly. After 33 weeks of POG the patient, gave birth by cesarean to a female with weight of 1,380 g, height of 40 cm and a heart rate of 48 bpm. The Apgar score was 6 at first minute and 7 at minute five, while the Silverman score was 3 at the first minute and 2 at minute five.

After birth, she was transferred to the Neonatal Intensive Care Unit (NICU). With ventilatory support phase II, nasal Continuous Positive Airway Pressure (CPAP). The patient was active and reactive to external stimuli. On the first day after birth, she still have CPAP support and a heart rate of 50 bpm without support of vasoactive amines (*Figure 1*), a radiography was taken as a control of the cardiopulmonary state (*Figure 2*). On the second day after birth, the patient remains bradycardic (60-65 bpm) with support of dobutamine, nonspecific human immunoglobulin and three doses of dexamethasone were added to the therapy.

On the third day after birth was placed a temporary external pacemaker, the electrodes were attached to the right ventricle (*Figure 3*),



**Figure 1.** Electrocardiographic trace of the newborn. A complete dissociation between auricular and ventricular rhythm is observed with an auricular beat rate of 140 and a ventricular beat rate of 50 bpm.



**Figure 2.** Control radiography taken at the first day after birth showing the normal findings in a newborn child.



**Figure 3.** Radiography taken after the placement of epicardial electrodes connected to the temporal pacemaker.

fixed to the skin and connected to an external generator, the procedure was performed by a pediatric cardiologist; omeprazole, cefotaxim and paracetamol were added to the management. Subsequently, the patient went to the (NICU) for surveillance, expecting a weight increase for the placement of a definitive pacemaker. On the fourth day, a 1-minute apnea episode was reported, a cephalic helmet was placed with oxygen at 35%, 3 L per minute and enteral stimulation was initiated. On the sixth day, the oxygen saturation was 93% with ambient air and weighted 1,100 g. A radiography with gastromegalia and distention of intestinal loops were reported. On the tenth day the cephalic helmet was retired. On the eleventh day, onfalocclisis were removed and was the last day of cefotaxime. At the next day the pacemaker is set to 125 bpm to improve her cardiac output.

Two months after surgery, the external pacemaker begins to fail, drops to 60 bpm and the pacemaker generator was replaced, the weight of the baby was of 2,730 g. A week later, she undergoes surgery. The external definitive pacemaker was placed and the heart rate is set at 100 bpm (Figure 4), it was supported with phase III ventilator with 42 cycles,  $\text{FiO}_2$  at 50%. Endopleural catheter was placed and removed the next day. One day after surgery,



**Figure 4.** Radiography taken after the implantation of the definitive pacemaker in retroperitoneal position.

the baby was extubated and a cephalic helmet was placed, the same was removed after three days, maintaining good oximetry.

The baby was discharged at 78 days due to clinical improvement with a weight of 3,150 g.

## DISCUSSION

The prevalence of congenital AV block it's found between 2 and 5% of fetuses and infants whose mothers are seropositive to ANA's,<sup>1</sup> who are usually diagnosed after the detection of AV block.<sup>13</sup> A prospective study which included 186 individuals, conformed by 146 serially screened fetuses with normal pregnancy outcomes, and 40 fetuses/neonates with a diagnosis of heart block or endocardial fibroelastosis, resulting that mothers of children with cardiac involvement had moderate to high anti-Ro/SSA antibody levels ( $\geq 50$  U/mL) as compared with only 44% of mothers of healthy infants ( $p < 0.0001$ ), and for fetuses with exposure to anti-Ro levels an odds ratio (OR) of 7.8 (range 0.4 to 159) was described.<sup>14</sup> Other studies have been reported with similar results, in which the recurrence rate of complete heart block (CHB) is at least two- to three-fold higher than the rate for a mother with anti-Ro/SSA and anti La/SSB antibodies.<sup>1,15-17</sup> In terms of prognostic a retrospective study in which they

included sixty fetuses with heart block of which thirty-two had complex cardiac malformations and 31 of them were associated with left isomerism. The outcomes of these fetuses were 22 terminations of pregnancy (TOP), three intrauterine fetal deaths (IUFD), three neonatal deaths (NND), two childhood deaths (CD) and only two survivors. They propose the fetal hydrops and the association with complex cardiac malformations as predictors of adverse outcome in cases of heart block where cases without cardiac malformations have a significantly better prognosis.<sup>18</sup> The diagnostic of congenital AV block is associated with high fetal and neonatal mortality, and independently whether the diagnosis is made before or after birth, both, undergo for the implantation of a pacemaker with earlier intervention and a significantly greater need for intervention among those diagnosed *in utero*.<sup>14</sup> With a mortality estimated around 8-16% in infants and half of this proportion in children and adults.<sup>13,14</sup>

For the fetal diagnosis of congenital AV block, the echocardiography has been determined as the gold standard, however, all M-mode and Doppler echocardiographic techniques that are based on the relationship between the atrial and ventricular mechanical events, can be well applied,<sup>19</sup> besides clinical application of fetal electrocardiography and fetal magnetocardiography are promising noninvasive and highly precise tools. 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,<sup>20</sup> 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. Coupled with this prenatal treatments may also include intravenous immunoglobulins and plasmapheresis, used alone or together in combination with steroids for the treatment of AV block<sup>21,22</sup> it could be considered in asymptomatic high degree AV blocks with specific risk conditions. Literature reports that two thirds of the affected pediatric population diagnosed with congenital heart block requires a pacemaker<sup>23</sup> in order to

prevent sudden cardiac death and to restore heart rate to avoid longstanding bradycardia.

We report a clear case of neonatal lupus syndrome according to the parameters of clinical, radiological, electrocardiographic diagnosis, the need of the implantation of a pacemaker was supplied, all the characteristics presented by the patient with a persistent bradycardia, made her fit for surgery. This procedure is presented as an excellent option for patients with structurally normal hearts instead of treatments with steroids of which there is great controversy around its use, however there is still discussion about whether the pacing should be permanent or temporary. Supporting the implementation of this procedure, there are reports of permanent pacemaker implantation in term and preterm newborns with very acceptable success and survival rates.<sup>24-26</sup> There is limited information about the success of implantation of pacemakers in neonates with AV block, therefore it is important to report the procedures performed and important points to achieve success of the surgery, although this success may be due to the pathophysiological aspects of each individual. The detection of more cases of this rare pathology, the refining of the diagnosis with the use of documented tools and the generation of devices adapted to the needs of the patient are integral factors for good care.

#### Acknowledgment

We thank CONACyT for the scholarship Number: 596911 for Lydia Enith Nava-Rivera and Dirección de Educación en Salud, Dirección General de Calidad y Educación en Salud de la Secretaría de Salud de México for the Social Service Research Grant to: Rodrigo Lozoya-Martínez, No. 4203610284.

**Conflict of interest:** The authors declare that no conflict interests exist.

#### REFERENCES

1. 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): 1658-1666.



2. Hunter LE, Simpson JM. Atrioventricular block during fetal life. *J Saudi Heart Assoc.* 2015; 27 (3): 164-178.
3. Ho SY, Esscher E, Anderson RH, Michaëlsson M. Anatomy of congenital complete heart block and relation to maternal anti-Ro antibodies. *Am J Cardiol.* 1986; 58 (3): 291-294.
4. Habash-Bseiso DE, Yale SH, Glurich I, Goldberg JW. Serologic testing in connective tissue diseases. *Clin Med Res.* 2005; 3 (3): 190-193.
5. Cozzani E, Drosera M, Gasparini G, Parodi A. Serology of Lupus Erythematosus: Correlation between Immunopathological Features and Clinical Aspects. *Autoimmune Dis.* 2014; 2014: 321359.
6. Maidhof W, Hilar O. Lupus: an overview of the disease and management options. *P T.* 2012; 37 (4): 240-249.
7. Gardiner HM, Belmar C, Pasquini L, Seale A, Thomas M, Dennes W et al. Fetal ECG: a novel predictor of atrioventricular block in anti-Ro positive pregnancies. *Heart.* 2007; 93 (11): 1454-1460.
8. Hunter LE, Simpson JM. Atrioventricular block during fetal life. *J Saudi Heart Assoc.* 2015; 27 (3): 164-178.
9. Friedman DM, Rupel A, Glickstein J, Buyon JP. Congenital heart block in neonatal lupus: the pediatric cardiologist's perspective. *Indian J Pediatr.* 2002; 69 (6): 517-522.
10. Buyon JP, Clancy RM, Friedman DM. Autoimmune associated congenital heart block: integration of clinical and research clues in the management of the maternal / foetal dyad at risk. *J Intern Med.* 2009; 265 (6): 653-662.
11. Dey M, Jose T, Shrivastava A, Wadhwa RD, Agarwal R, Nair V. Complete congenital foetal heart block: a case report. *Facts Views Vis Obgyn.* 2014; 6 (1): 39-42.
12. Puri S, Pooni P, Mohan B, Bindal V, Verma S, Verma S et al. Pregnancy with SLE and fetal congenital heart block: a case report. *Cardiol Res.* 2013; 4 (3): 126-128.
13. Baruteau AE, Pass RH, Thambo JB, Behaghel A, Le Pennec S, Perdreau E et al. Congenital and childhood atrioventricular blocks: pathophysiology and contemporary management. *Eur J Pediatr.* 2016; 175 (9): 1235-1248.
14. Jaeggi ET, Hamilton RM, Silverman ED, Zamora SA, Hornberger LK. Outcome of children with fetal, neonatal or childhood diagnosis of isolated congenital atrioventricular block. A single institution's experience of 30 years. *J Am Coll Cardiol.* 2002; 39 (1): 130-137.
15. Martínez-Sánchez N, Pérez-Pinto S, Robles-Marhuenda A, Arnalich-Fernández F, Martín-Cameán M, Hueso-Zalvide E et al. Obstetric and perinatal outcome in anti-Ro/SSA-positive pregnant women: a prospective cohort study. *Immunol Res.* 2017; 65 (2): 487-494.
16. Eronen M, Miettinen A, Walle TK, Chan EK, Julkunen H. Relationship of maternal autoimmune response to clinical manifestations in children with congenital complete heart block. *Acta Paediatr.* 2004; 93 (6): 803-809.
17. Ambrosi A, Salomonsson S, Eliasson H, Zeffer E, Skog A, Dzikaite V et al. Development of heart block in children of SSA/SSB-autoantibody-positive women is associated with maternal age and displays a season-of-birth pattern. *Ann Rheum Dis.* 2012; 71 (3): 334-340.
18. 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 Gynecol.* 2005; 26 (1): 4-15.
19. Fouron JC, Proulx F, Miró J, Gosselin J. Doppler and M-mode ultrasonography to time fetal atrial and ventricular contractions. *Obstet Gynecol.* 2000; 96 (5 Pt 1): 732-736.
20. Rein AJ, Mevorach D, Perles Z, Gavri S, Nadjari M, Nir A et al. Early diagnosis and treatment of atrioventricular block in the fetus exposed to maternal anti-SSA/Ro-SSB/La antibodies: a prospective, observational, fetal kinetocardiogram-based study. *Circulation.* 2009; 119 (14): 1867-1872.
21. Friedman DM, Llanos C, Izmirly PM, Brock B, Byron J, Copel J et al. Evaluation of fetuses in a study of intravenous immunoglobulin as preventive therapy for congenital heart block: Results of a multicenter, prospective, open-label clinical trial. *Arthritis Rheum.* 2010; 62 (4): 1138-1146.
22. Ruffatti A, Milanesi O, Chiandetti L, Cerutti A, Gervasi MT, De Silvestro G et al. A combination therapy to treat second-degree anti-Ro/La-related congenital heart block: a strategy to avoid stable third-degree heart block? *Lupus.* 2012; 21 (6): 666-671.
23. Lazzerini PE, Capecechi PL, Guideri F, Acampa M, Galeazzi M, Laghi Pasini F. Connective tissue diseases and cardiac rhythm disorders: an overview. *Autoimmun Rev.* 2006; 5 (5): 306-313.
24. Welch EM, Hannan RL, DeCampi WM, Rossi AF, Fishberger SB, Zabinsky JA et al. Urgent permanent pacemaker implantation in critically ill preterm infants. *Ann Thorac Surg.* 2010; 90 (1): 274-276.
25. Takabayashi S, Ito H, Shimpo H, Sawada H, Mitani Y, Komada Y. Emergent permanent pacemaker implantation in a premature 1,502 g neonate. *Jpn J Thorac Cardiovasc Surg.* 2005; 53 (4): 199-201.
26. Tomita Y, Imoto Y, Tominaga R, Yasui H. Successful implantation of a bipolar epicardial lead and an autocapture pacemaker in a low-body-weight infant with congenital atrioventricular block: report of a case. *Surg Today.* 2000; 30 (6): 555-557.

#### Correspondence to:

#### Dr. Javier Morán Martínez

Departamento de Biología  
Celular y Ultraestructura,  
Centro de Investigación Biomédica,  
Facultad de Medicina,  
Universidad Autónoma  
de Coahuila Unidad Torreón.  
Gregorio A. García Núm. 198 sur,  
27000, Torreón, Coahuila, México.  
Oficina: +52 87 17171077  
E-mail: javmoran@yahoo.com