

Treatment of Congenital Complete Atrioventricular Heart Block With Permanent Epicardial Pacemaker in Neonatal Lupus Syndrome

Sema Tanriverdi,^{1*} Zual Ulger,² Betul Siyah Bilgin,¹ Nilgun Kultursay,¹ Mehmet Yalaz,¹ Yuksel Atay,³ and Ozge Altun Koroglu¹

¹Department of Pediatrics, Ege University, Faculty of Medicine, Division of Neonatology, Izmir, Turkey

²Department of Pediatrics, Ege University, Faculty of Medicine, Division of Pediatric Cardiology, Izmir, Turkey

³Department of Cardiovascular Surgery, Ege University, Faculty of Medicine, Izmir, Turkey

*Corresponding Author: Sema Tanriverdi, Department of Pediatrics, Ege University, Faculty of Medicine, Division of Neonatology, Izmir, Turkey. Tel: +90-2323901306, Fax: +90-2323901011, E-mail: drsemarala@yahoo.com

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Introduction: Neonatal lupus syndrome (NLS) is a passively acquired autoimmune condition due to the transplacental passage of maternal anti-Ro/SSA and anti-La/SSB antibodies in mothers with systemic lupus erythematosus (SLE), and congenital complete heart block (CHB) is its most serious manifestation. Skin and hepatic involvement may occur in later infancy.

Case Presentation: A term infant with fetal bradycardia, detected at the 23rd gestational age, was diagnosed with CHB due to NLS and was successfully treated with a permanent epicardial pacemaker. The patient was reported here due to rarity of the procedure in neonatal period.

Conclusions: Mothers with SLE should be screened and closely followed up during pregnancy for the development of fetal atrioventricular (AV) block.

Keywords: Heart Block; Neonatal Lupus Syndrome; Artificial Cardiac Pacemaker; Therapy

1. Introduction

Neonatal lupus syndrome (NLS) is a passively-acquired autoimmune condition due to the transplacental passage of maternal anti-Ro/SSA and anti-La/SSB antibodies in infants of mothers with systemic lupus erythematosus (SLE). Congenital complete heart block (CHB) is the most serious manifestation of NLS (1). Although the pathogenesis is not clarified, it is suspected that bindings of anti-Ro/SSA and/or anti-La/SSB antibodies to fetal cardiac tissue result in heart block, leading to autoimmune injury of the atrioventricular node and the surrounding tissue. The most vulnerable period for the fetus is during the 18-24 gestational weeks and CHB may present with fetal bradycardia during this period (2).

Weekly ultrasounds, to follow the fetal heart rate (FHR) pattern were performed and FHR was stable at 52 - 56 beats/minute with no features of hydrops.

2. Case Presentation

A female term infant with 2870 g birth weight was born to a 29-year-old mother with SLE at 37-38 weeks gestational age by C-section. Fetal bradycardia was diagnosed at the 23rd week of gestation at another center and the mother applied to our obstetrics department at the 27th week. The

infant was hospitalized in the neonatal intensive care unit in Izmir, Turkey, in 2012 because of fetal bradycardia in the fetal ultrasound and postnatal bradycardia in the electrocardiogram. The heart rate was 55/minute with arrhythmia with normal respiratory rate (50/minute), arterial blood pressures (79/45 mmHg), and arterial oxygen saturation (98%). The variables of heart rate were measured with electrocardiogram in Philips IntelliVue MP40 and MP50 (Philips Medical Systems, BG Eindhoven, The Netherlands). A 2/6° systolic murmur was heard. The remaining physical examinations and hematologic and biochemical parameters were all normal. The white blood cell (WBC) count: 11300 mm³, (normal range: 9,000 - 30,000), hemoglobin: 15 g/dL (normal range: 13.5 - 22), hematocrit: 43.7% (normal range: 42-60), platelet count: 248 000 mm³ (normal range: 150 000 - 350 000), urea: 14 mg/dL (normal range: 16.6 - 48.5), creatinine: 0.6 mg/dL (normal range: 0.24 - 0.85), blood glucose: 83 mg/dL (normal range: 50 - 80), aspartate aminotransferase (AST): 37 U/L (normal range: 5 - 40), alanine aminotransferase (ALT): 30 U/L (normal range: 5-40), creatine kinase (CK): 180 U/L (normal range: 20 - 200), and CK myocardial b fraction (MB): 20 U/L (normal range: 3 - 25).

The patient was monitored and complete heart block was detected in the electrocardiogram (Figure 1 A). He-

modynamically insignificant patent ductus arteriosus (PDA) (2 mm) and patent foramen ovale (PFO) (2 mm) were detected on echocardiography.

The anti-Ro/SSA and anti-La/SSB levels of the infant were 138 U/mL and 102 U/mL, respectively (normal range < 15 U/mL for both). The clinical findings stayed stable while the heart rate was between 45 - 60/minute. At day 28, a permanent epicardial pacemaker was placed and the rate of the generator was adjusted as 110/minute (Figure 1 B). The patient was discharged at day 35. The heart rate of the generator was calibrated

By the biomedical engineering staff with regular controls. Any complication was observed.

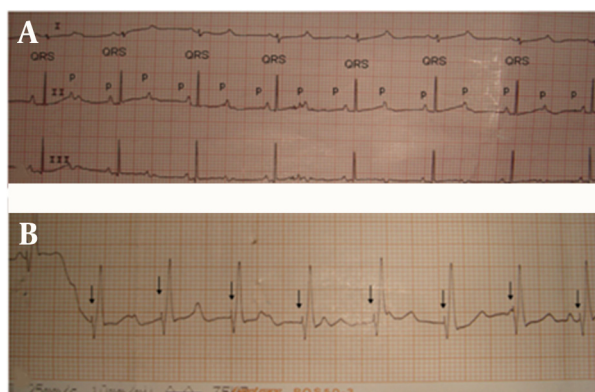


Figure 1. Complete Heart Block With Irregular Atrial and Ventricular Rates (A) and Normal Electrocardiogram After Pacemaker Placement (B)

3. Discussion

NLS is a passively acquired autoimmune condition with cutaneous erythematous lesions, congenital atrioventricular block, liver dysfunction, and hematological problems, due to transplacental passage of maternal anti-Ro/SSA and anti-La/SSB1 antibodies. The most serious manifestation of NLS is CHB which presents with fetal bradycardia between 18 and 28 weeks of gestation. First or second degree heart block detected in utero or at birth can progress to CHB, which is usually irreversible (3).

The management of CHB includes steroid and IVIG treatments in early weeks of gestation (before the 16th week), since cardiac findings usually occur between 18 - 24 gestational weeks (4). However, prophylactic steroid therapy is not routinely recommended, since the occurrence of CHB is between 1% - 2% in infants of mothers with SLE as well as the possibility of neurodevelopmental problems due to steroids in the fetus. It may be prescribed to pregnant women with antibodies or a history of previously delivered infants with CHB when fetal first degree atrioventricular (AV) block is diagnosed to prevent its progress to CHB. The potential mechanisms of IVIG in preventing the tissue damage are increased elimination of maternal anti-Ro/SSA and anti-La/SSB antibodies, decreased transplacental transport of antibodies, and modulation of

inhibitory signaling on macrophages with consequent reduction of inflammatory response and fibrosis (3-7).

Neonatal successful permanent pacemaker implantation is rarely reported in patients with CHB (5-7). The indications for permanent pacemaker implantation in infants are ventricular rate of < 55 beats/minute, atrial rate of > 140 beats/minute, wide QRS complexes, congenital heart disease, heart failure, and prolonged QT. Patients requiring permanent pacemaker implantation have high mortality rates due to dilated cardiomyopathy in long term (8). Permanent pacemaker was placed at day 28 in our patient since she had a heart rate of 45 - 50 beat/minute. The heart rate was 110/minute after the pacemaker implantation and she is on follow-up program for possible late-onset problems such as dilated cardiomyopathy, congestive heart failure, and hepatic and skin involvement of NLS.

The successful treatment of CHB with a permanent epicardial pacemaker as in our case is very rarely reported in the neonatal period. Mothers with SLE or with positive anti-Ro/SSA and anti-La/SSB antibodies should be screened and closely followed up during pregnancy for the development of fetal AV block since prevention of CHB may be possible in the early period of gestation.

Authors' Contributions

Sema Tanriverdi prepared the manuscript and finally revised it. Zulal Ulger, Betül Siyah Bilgin, and Yuksel Atay helped in follow up of the patient and acquisition of data. Mehmet Yalaz developed the original idea, revised the manuscript, and supervised the treatment. Ozge Altun Koroglu and Nilgun Kultursay helped in follow up of the patient, acquisition of data, abstracted the findings, and revised the manuscript.

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