Long-Term Follow-Up of Children with Heart Block Born from Mothers with Systemic Lupus Erythematosus: A Retrospective Study from the Database Pediatric and Congenital Heart Disease in University Hospitals Leuven

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**Background:** Children from mothers with systemic lupus erythematosus are frequently born with congenital heart block. This study aimed at evaluating long-term outcome because long-term data are scarce.

**Methods:** In the database of pediatric and congenital heart disease (University Hospitals Leuven), 19 children from systemic lupus erythematosus mothers and who were born with or developed atrioventricular block were identified. All records were reviewed for disease course and outcome.

**Results:** Median follow-up time was 7 years (interquartile ranges [IQR] 4.5–13 years). One child had no heart block at birth and developed only a first-degree block during follow-up. One had a second-degree heart block and developed a complete heart block. Seventeen patients (89%) were born with a complete heart block. Seventeen patients (89%) needed a definitive pacemaker. In all, epicardial leads were used at first implantation. Eighty-two percent received their pacemaker in the first year of life. The first battery had a median lifetime of 5 years (IQR 3.5–5 years), the second 6 years (IQR 4.5–6.3 years), and the third 5 years (IQR 5–6 years). Note that 47% of patients needed a lead replacement due to lead problems. Only one pericardial tamponade after pacemaker implantation. No device or lead infections occurred. The left ventricular systolic function at latest follow-up was normal for all. No patients died.

**Conclusion:** In children with heart block born from systemic lupus erythematosus mothers, an early need for pacemaker implantation was documented. The overall battery life was acceptable, but there was a high need for lead replacement. Complication rate was low. Late outcome was good. (PACE 2016; 00:1–9)

**Introduction**

Pregnant patients with systemic lupus erythematosus are known to be at increased risk for miscarriage, intrauterine growth restriction, preterm delivery, and perinatal fetal death.¹ Much has been reported concerning the outcome of mothers with systemic lupus erythematosus during pregnancy. However, literature is scarce about the outcome of newborns from systemic lupus erythematosus mothers suffering from heart block.

Congenital atrioventricular heart block is a rare illness, affecting one in 15,000–20,000 live births.² The large majority of congenital heart block cases (60–90%) presenting in utero or in the neonatal period are due to neonatal lupus. The fetal heart is perfused with maternal anti-Ro/anti-Sjögren’s syndrome-related antigen antibodies crossing the placenta.³,⁴ Congenital heart block is a rare manifestation among neonatal lupus symptoms and the prevalence is now reported to be 1–2% in pregnant women with anti-SSA antibodies.⁵

Maternal antibodies, which start crossing the placenta as early as 11 weeks of gestation, are associated with the development of cardiac...
anomalies, rash, and/or various liver and blood cell abnormalities in the newborn. Clearance of the antibodies occurs in the 6th to 8th month of postnatal life. In contrast to regenerative processes in the skin, the hepatobiliary, and the hematologic system, this regeneration does not occur in cardiac tissue. Indeed, definitive reversal of a third-degree congenital heart block has never been observed.\textsuperscript{6} This is explained by disruption of the atrioventricular conduction system by the maternal antibodies. The latter cause inflammation with subsequent fibrosis and calcification, which leads to an irreversible complete atrioventricular block.\textsuperscript{6}

In 1998, Buyon et al. published the first large series of autoimmune-associated congenital heart block based on “The Research Registry for Neonatal Lupus” in the United States. They found that autoantibody-associated congenital heart block is not coincident with major structural abnormalities, carries a substantial mortality in the neonatal period, and frequently requires pacing.\textsuperscript{7} The aim of our study was to evaluate children with congenital heart block born from mothers with systemic lupus erythematosus evaluating (1) the presence at birth and persistence during follow-up of heart block, (2) the presence of associated structural cardiac abnormalities at birth and during follow-up, (3) the need for and timing of complications related to pacemaker implantation, (4) late effect on left ventricular systolic function, and (5) survival.

Materials and Methods

Study Population

All patients were identified through the database of pediatric and congenital heart disease from the University Hospitals Leuven, updated until April 2014. The inclusion criteria for this study were: (1) born with or developed congenital heart block and (2) born from a mother with systemic lupus erythematosus. The study protocol was reviewed and approved by the Ethics Committee of the University Hospitals Leuven.

The search based on the key words of the inclusion criteria resulted in a list of 24 files. Five files had to be excluded because these patients did not meet the inclusion criteria, as these patients were born from a mother in whom the diagnosis of systemic lupus erythematosus was not confirmed.

Variables

All patient files were reviewed for the presence at birth, development of, and degree of congenital heart block; the need for and timing of pacemaker implantation; possible complications; and death. Echocardiography at first presentation and at latest follow-up was reviewed for associated structural cardiac lesions and/or evaluation of left ventricular function. Moderate valvular insufficiency was noted when it was graded at least 2 of 4. Mild valvular insufficiency was noted when it was graded less than 2 of 4. Neonatal myocarditis was defined as the presence of biventricular dysfunction and elevated cardiac enzymes.

Statistical Analysis

Mainly descriptive statistics were used. Because of the low number of patients, continuous variables were reported by median and interquartile ranges (IQR). Proportions were mentioned by numbers and percentages. To identify differences in consecutive battery survival, Kaplan-Meier analysis and log rank testing were done. Statistical analyses were performed in Excel (Microsoft Office 2013, Microsoft Corp., Redmond, WA, USA) and by using Statistical Package for the Social Sciences (version 20; IBM Corp., Armonk, NY, USA). A P value < 0.05 was considered to be statistically significant.

Results

Baseline Characteristics

Demographics

The majority of the patients were female (68%, 13/19). Seventy-four percent of the patients (14/19) were from Caucasian origin.

The birth weight and gestational age at birth were missing in four files. The median birth weight was 2.345 kg (IQR 1.865–2.237 kg) and the median gestational age was 34 weeks (IQR 32.25–36.75 weeks). Some deliveries were induced early because of progressing heart block, threatening heart failure in the fetus, fetal hydrops, or fetal distress caused by neonatal lupus.

Immune Status of the Mothers

Eighty-four percent (16/19) of mothers had a diagnosis of pure systemic lupus erythematosus, 11% (2/19) had a diagnosis of mixed systemic lupus erythematosus with Sjögren’s syndrome, and 5% (1/19) had a pure Sjögren’s syndrome.

We tried to identify the specific immune status of the mothers but in 16% (3/19) of files, the immune status could not be identified because it was not mentioned in their children’s file and we were not able to get into contact with the mothers or their treating physician. All the remaining 16 mothers (100%) tested positive for anti-Rho/SSA antibodies; 63% (10/16) tested positive for anti-La/SSB antibodies.

Prenatal and Postnatal Therapy

In 52% (10/19) of patients, corticosteroids were administered before birth. In 16% (3/19)
of patients, corticosteroids were administered after birth. In one patient (1/19), terbutaline was given before birth. In none of our patients, intravenous immunoglobulin therapy was given or plasmapheresis applied.

Heart Block at Birth and during Follow-Up

The median follow-up time was 7 years (IQR 4.5–13 years). Only one patient (1/19, 5%) had no heart block at birth, but developed a first-degree atioventricular block as a neonate with a PR interval of 224 ms. One patient (1/19, 5%) had a second-degree block, but developed a complete heart block later on. All the other patients (17/19, 89%) were born with a complete heart block.

Associated Congenital and Structural Heart Disease at Birth

In 58% of the patients (11/19), there was some kind of structural heart defect. Atrial septal defects (ASDs; 4/19, 21%), ventricular septal defects (VSDs; 3/19, 16%), and persistent ductus arteriosus (PDA; 6/19, 32%) were noted. One patient had a monocoronary system (1/19, 5%). Twenty-one percent of patients (4/19) had a moderate tricuspid insufficiency. One patient (1/19, 5%) had a moderate mitral insufficiency. There were patients with prominent right ventricular hypertrophy (2/19, 11%) and left ventricular hypertrophy (3/19, 16%). In six patients (6/19, 32%), there was a significant dilatation of the left ventricle, however, with normal function. Neonatal myocarditis was diagnosed in three patients (16%, 3/19).

Pacing

Temporary Pacing

Before the definitive pacemaker implant, six patients (32%) needed temporary pacing with epicardial temporary leads and an external pacing system as a dedicated temporary pacing method. Two (11%) were bridged with transesophageal leads before the epicardial system was implanted. Both patients received their transesophageal pacing in a referral center, but detailed information about the technique could not be obtained. One patient was paced for 20 days; the duration of transesophageal pacing in the other patient is not known.

The reasons for temporary pacing were cardiogenic shock (17%), fetal hydrops (17%), or very slow ventricular escape rhythm with bradycardia <70/min (67%).

Definitive Pacemaker Implantation

The patient with first-degree heart block did not progress to a higher degree heart block and did not undergo pacemaker implantation at the current age of 21 years. One patient received epicardial leads, but had a sufficient escape rhythm, so that no pacemaker was implanted until today. All the other patients (17/19, 89%) received a definitive pacemaker, all with epicardial leads at first. In Figure 1, the timing of the first
pacemaker implantation is visualized indicating that after the first year, 82% of patients have received a pacemaker. Forty-one percent (7/17) of pacemakers were implanted during the first month of life. The earliest pacemaker implantation was done on the first day of life; the latest pacemaker implantation was done at the age of 7 years.

**Complications Immediately Related to Pacemaker Implantation**

One of the two patients who needed temporary transesophageal pacing developed an esophageal stenosis. This was the patient who received 20 days of transesophageal pacing. In the immediate postpacemaker and lead implantation period were four patients (4/17, 24%) in whom pericardial effusion was detected on routine echocardiography. One of them (1/17, 6%) had signs of pericardial tamponade and needed two repetitive punctures to drain the pericardial fluid. No other complications were found; more specifically, there were no reports of device or lead infections.

**Battery Replacement**

Two patients were lost to follow-up. The remaining 15 patients had one or more battery replacements. The patients received their first battery replacement after a median time of 5 years (IQR 3.5–5 years). Eight patients (53%) needed a second battery replacement, after their second battery had lasted for a median time of 6 years (IQR 4.5–6.3 years). Three patients (20%) received a third battery. Figure 2 is a Kaplan-Meier plot where log rank testing was performed to identify differences between battery life of the first, the second, and the third battery.

Eight patients (53%) had a dual-chamber device implanted when battery replacement was needed: in five patients (33%) the second ventricular lead was used as a backup, two patients (13%) were paced in both ventricles because they had no underlying escape rhythm, and one patient (7%) received dual right ventricular pacing because a slow ventricular escape rhythm around 30 beats/min.

**Leads**

All patients (17/17) received an epicardial pacing system at first. All leads were Medtronic Capsure Epi model 4965 steroid eluting leads (Medtronic, Minneapolis, MN, USA). Two-thirds of the patient population (71%, 12/17) were paced on the right ventricle. Three patients (18%) were paced on the left ventricle and two patients (12%) were paced on both ventricles.

During follow-up, a total of 10 patients (59%) underwent lead replacement after a median time...
Table I. Listing of the Temporary Pacing Mode, Number, and Location of Leads and Pacing Mode at Implantation and Last Follow-Up for Each Patient in the Cohort

<table>
<thead>
<tr>
<th>Temporary Leads at Transesophageal Epicardial Leads at Implantation</th>
<th>Pacing Mode at Implantation</th>
<th>Leads at Latest Follow-Up</th>
<th>Pacing Mode at Latest Follow-Up</th>
</tr>
</thead>
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<tr>
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<td>0 2 0</td>
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<tr>
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<td>Yes</td>
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<tr>
<td>Patient 17</td>
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</tbody>
</table>

LV = left ventricle; RV = right ventricle.

of 5 years (IQR 4–5 years). There were multiple reasons found for the lead replacements: (1) six patients (35%) had a lead fracture; two of them even had two fractures, (2) two patients (12%) were diagnosed with lead dysfunction (high threshold without lead fracture), (3) two patients (12%) received an elective lead replacement during the procedure for battery replacement with upgrade to a dual-chamber device. Table I shows an overview of the number of leads at implantation and at latest follow-up.

**Pacing Mode at Implantation and Electrical Analysis at Latest Follow-Up**

The majority of the patients (14/17, 82%) were programmed in VVI mode at first; only two of 17 patients (11%) were immediately programmed in DDD mode. From one patient no information concerning the pacing mode at first programming was found.

The pacing mode after implantation and at latest follow-up is summarized in Table I. A minority of the patients were still programmed in a VVI or VVI-R mode (5/17, 29%). There were 12 patients (71%) programmed in DDD, DDD-R, or cardiac resynchronization therapy mode. Nine of them (75%) received implantation of an atrial lead during follow-up and three of them (33%) received the atrial lead at the first lead implantation procedure, but in one patient the atrial lead was initially not used. The majority of the patients programmed in DDD/DDD-R mode had a cardiac resynchronization therapy device (7/9, 78%), where the second ventricular lead was used as a backup but not functional. Three patients (18%) were programmed in biventricular pacing mode for safety at latest follow-up; two (12%) of them were programmed in cardiac resynchronization therapy biventricular pacing mode because they had no ventricular escape rhythm, one of them (6%) had dual right ventricular pacing because of a very slow ventricular escape rhythm around 30 beats/min.

**Left Ventricular Systolic Function and Structural Heart Disease during Follow-Up**

At birth, 26% of the patients (6/19) had some degree of decreased left ventricular ejection fraction. The echocardiography at latest follow-up was reviewed in all patients. For evaluation of the left ventricular function some records used an ejection fraction, while other records used fractional shortening. One patient (5%) had a borderline left ventricular systolic function...
due to septal dyskinesia. All the other patients had a normal left ventricular systolic function at latest follow-up. Two patients (11%) had a cardiac resynchronization therapy device with biventricular pacing mode. Indication for implantation of the biventricular device was not a decreased left ventricular function, but these patients had no escape rhythm and needed 100% pacing.

The ASD persisted in all four patients (100%). Two patients (2/4, 50%) underwent a surgical repair and one (1/4, 25%) received an ASD closure device. In one patient (1/4, 25%), the ASD persisted and was not corrected during follow-up. All three VSDs (100%) closed spontaneously. Of the PDA, 67% (4/6) closed spontaneously, one (1/6, 17%) was closed with a Rashkind umbrella, and one (1/6, 17%) was surgically closed.

The presence of moderate insufficiency of the mitral and tricuspid valve was in none of the patients graded higher than a mild insufficiency at latest follow-up. The left and right ventricular hypertrophy that was present at birth normalized in all patients (100%) during follow-up. Left ventricular dilation (present in 6/19 patients at birth) persisted in four of six patients (67%); in three of them (3/6, 50%) the dilation was only graded mild, in one (1/6, 17%) the dilation was graded moderate.

Survival during Follow-Up

None of our patients died.

Discussion

Nineteen children with congenital heart block born from mothers with systemic lupus erythematosus were identified through the database of pediatric and congenital heart disease in University Hospitals Leuven. Eighty-nine percent of patients presented immediately with a complete heart block at birth. After a median follow-up time of 7 years (IQR 4.5–13 years), 95% of patients had a complete heart block. Eighty-nine percent of patients needed a definitive pacemaker implantation, all with surgically placed epicardial leads at first. Complication rate immediately related to pacemaker implantation was low with only one pericardial tamponade. A high percentage of lead replacements in 59% of our patient population was noted. In 58% of patients, associated structural heart disease was present, and 16% of patients had a neonatal myocarditis. The left ventricular systolic function a latest follow-up was normal in all patients.

Demographics

The risks of premature and low weight baby rates are increased in mothers with systemic lupus erythematosus. A recent Chinese study of 41 patients found a mean gestational age of 37.6 ± 3.3 weeks and a mean birth weight of 2.510 ± 0.757 kg. In our study, the median birth weight and gestational age were considerably lower than in the higher mentioned study. This can be explained by the fact that some deliveries were induced early because of progressing heart block, threatening heart failure in the fetus, fetal hydrops, or fetal distress.

Structural Heart Disease

Structural congenital heart disease has been reported in 16–42% of patients with congenital heart block. We found structural heart disease in 58% of the cases. It has been suggested that cardiomyopathy may also be a separate manifestation of neonatal lupus. Neonatal myocarditis, for example, is a manifestation of myocardial infiltration by the maternal antibodies and we found 16% of patients with myocarditis. Nevertheless we noticed a normal left ventricular function at latest follow-up in all patients, although in literature the outcome of neonatal lupus-associated myocarditis is very poor, with more than 80% of patients needing a heart transplant or dying. One of the three patients with neonatal myocarditis (33%) received prenatal therapy with corticosteroids, one of the three patients (33%) received postnatal therapy with corticosteroids, and one (33%) received no therapy.

We cannot exclude that there could be a selection bias because our study started from the database of pediatric and congenital cardiology—while most studies are based on systemic lupus erythematosus registries. This could explain for the higher prevalence of structural heart disease in our population.

Pacing

Battery Survival

We describe an early timing of pacemaker implantation with the majority of pacemakers implanted during the first year of life (75%) and even 40% during the first month. Battery life is known to be relatively shorter in neonates compared with children and adults because of the fast heart rate and complete pacemaker dependency. Also in newborns, the smallest battery possible is implanted. A retrospective study in 22 neonates who underwent epicardial pacemaker implantation because of congenital heart block or sinus node dysfunction reported a median battery life of 4.1 years, which is little shorter than the median time of 5 years for the first battery replacement in our analysis.
Complication Rate Related to Pacemaker Implantation

One of the two patients who was paced transesophageally for 20 days developed an esophageal stenosis and needed multiple esophageal dilations afterwards. We want to stress this complication as a warning. In the past, this procedure was used in our center in extremely preterm infants, but after a few days the electrodes showed electrolysis and infants developed caustic esophageal stenosis. Nowadays we use temporary epicardial pacing leads with an external pacemaker system.

Despite the early timing of pacemaker implantation, we found a surprisingly low complication rate: there was only one patient with a cardiac tamponade with the need for pericardiocentesis. More important, there were no device infections, which is one of the most feared complications with a high morbidity. No fatal complications occurred.

Leads

All of our patients (17/17) received an epicardial pacing system at first. Taken into consideration the very early need for pacemaker and lead implantation, this is not an abnormal finding. Indeed, inserting a lead by the transvenous approach in a small sized vessel carries the inherent risk of stenosis or thrombosis.

A total of 10 patients (59%) needed a lead replacement due to lead fracture (35%), lead failure due to high pacing thresholds (12%), or during an elective procedure (upgrade to dual-chamber device; 12%). Reoperations are an inevitable problem associated with pediatric pacemaker therapy and a very important predictor for lead problems is the age of the patients at implantation of the leads,12,13 which was very young in our population. Compared to a study in 93 patients with congenital heart disease having epicardial leads,13 we found a higher percentage of lead failures, but the median time period before failure was comparable. In comparison to a retrospective study looking at long-term outcome of epicardial leads, 38% had a lead problem after 5 years, which is a lower percentage than in our findings.14 We have to mention that the cause of lead failure in one patient was a twist in the pacemaker battery caused by a fall while riding her bicycle. There could also be a sample size problem as our patient group was very small. In comparison to a retrospective study in Italy, where the mean age of implantation was significantly older, similar results were found: 53% of lead failure in pediatric patients with an epicardial system.15

Pacing Mode

Almost all of the studied patients were programmed in a VVI mode at first, but at latest follow-up almost all of our patients were paced in the atrium and the ventricle. The contribution of atrial contraction to cardiac output has been the subject of extensive research. A small study published in 2011 found a noninvasive method to quantitatively measure the cardiac output in VVI versus DDD mode (with optimized atrioventricular delays) in patients with a congenital heart block. They found a significant increase of the cardiac output of 18% (P < 0.001) when patients were programmed from right ventricular pacing only to atrioventricular synchronous pacing.16 This supports the hemodynamic advantage of atroventricular synchronous pacing and therefore the implantation of a two-chamber pacing system with programming in DDD mode. One patient received an atrial lead at implantation, but was initially programmed in VVI mode despite having both an atrial and a ventricular lead. It is unclear to us in the patient file why this was done.

The majority of our patients had two ventricular leads, where the second ventricular lead was used as a backup but not functional. This approach was useful as in 29% (2/7) of patients with a lead problem the second lead had to be used. Two patients, who had no underlying escape rhythm, were paced in both ventricles (cardiac resynchronization therapy), although they had a preserved ejection fraction before implantation of the cardiac resynchronization therapy device.

There is evidence that in patients with bradycardia and need for pacing, there is superiority of biventricular pacing to right ventricular apical pacing in the prevention of left ventricular adverse remodeling and deterioration of systolic function, even in patients with preserved ejection fraction. Because these patients had no escape rhythm and needed 100% pacing, a biventricular pacing device was chosen to prevent deterioration of the left ventricular function.17

Left Ventricular Systolic Function

Gebauer and Tomek evaluated the systolic left ventricular function of 82 pediatric patients with complete heart block who had been 100% paced on the right ventricle.18 The incidence of left ventricular dilatation or dysfunction was 13% (11/84) and epicardial right ventricular free wall pacing was the only independent risk factor. In our population, all had a normal left ventricular ejection fraction at latest follow-up. Two patients had biventricular pacing, not because a decreased left ventricular function, but because they had no ventricular escape rhythm. For this reason pacing on two leads was chosen for safety. As we described earlier in the article, there is some evidence that biventricular pacing can prevent adverse remodeling of the left ventricle, even in patients with preserved ejection fraction.18 There
was one patient with borderline left ventricular systolic function at latest follow-up, but this was the patient who had had an epicardial lead implantation, but no pacemaker because of a sufficiently high escape rhythm.

A recent study confirmed that the site of ventricular pacing has a major impact on left ventricular mechanical synchrony, efficiency, and pump function in children who require lifelong pacing. Of the sites studied, left ventricle apex/left ventricle midlateral wall pacing has the greatest potential to prevent pacing-induced reduction of cardiac pump function. Nevertheless the majority of patients in our population were paced on the right ventricle without complications.

Survival
The mortality in children with congenital heart block is estimated in studies at 8–16% and even higher up to 29% in children with associated cardiomyopathy. We found no mortality in our group, which is a very favorable outcome. This supports the need for and long-term efficacy of early pacemaker implantation in patients with congenital heart block—especially because the natural course of congenital heart block is known to have a high mortality.

Limitations
Our study is retrospective starting from the database of pediatric and congenital heart disease with a known risk for selection bias. Because this is a database of congenital cardiology, we are not aware of fetal losses and there might be a selection bias affecting our survival rate.

It is a relatively small study from a single center. We decided to use no comparison group of paced infants without autoimmune disease in the mother as the low numbers would have insufficient statistical power to run valuable comparative statistics. Also, our patient database was inhomogeneous concerning patient age and time of follow-up. Some patients were lost at follow-up.

We had not enough relevant information concerning the immune status of the newborns as anti-Rho/SSA and anti-La/SSB status was not taken. Data from referral centers were difficult to obtain, as in the both patients who received temporary transeosophageal pacing, detailed information about the technique used was missing.

Conclusions
This is a retrospective study with a cardiologic viewpoint looking at fetal outcome and the need for pacemaker implantation in newborns with congenital heart block from systemic lupus erythematosus mothers.

A very early timing of pacemaker implantation after birth seems needed with very few complications and an acceptable battery life. The largest problem remained frequent lead problems with 47% of patients needing lead replacement because of lead fracture or lead failure after 5 years.

Systolic left ventricular function was preserved at latest follow-up despite pacing. A lifelong follow-up in these patients is indicated, not only because of the need for electrical follow-up after pacemaker implantation, but also because of the high prevalence of associated structural heart disease.

References


