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Successful perinatal management and pacemaker stimulation during the first hour of life in a 1.6 kg newborn with autoimmune congenital complete heart block diagnosed prenatally

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Autoimmune congenital complete heart block (CCHB) occurs in 2–5% of pregnancies with anti-Ro/SSA (most common) and/or anti-La/SSB positive antibodies. The risk is higher in women with anti-Ro antiobodies in moderate (≥ 50 U/mL) and high (> 100 U/mL) titers, whereas an anti-La high titer is associated with non-cardiac features of neonatal lupus. After 16 weeks of gestation, antibodies cross the placenta and may destroy cardiomyocytes and conductive tissue in the atrio-ventricular node causing complete third degree heart block in more than 80% of cases. The highest risk of block development occurs up to 28 weeks of gestation. Current management of the condition includes: 1) decreasing inflammation through the administration of maternal fluorinated steroids and/or plasmapheresis; 2) increasing fetal cardiac output through beta-agonists administration; and 3) digoxin and/or lasix to treat hydrops and ventricular dysfunction. Direct fetal pacing was also tried but without success. Preliminary data suggest that a prophylactic treatment with hydroxychloroquine may be beneficial in preventing CCHB, but the safety of this drug should be evaluated. However, these therapies only have limited benefits and the mortality rate due to autoimmune CCHB is 16–30% of which 70% die in utero. Antibody-associated myocardial inflammation, dilated cardiomyopathy, ventricular rate < 55 bpm, impaired left ventricular function, fetal hydrops, diagnosis of CHB< 20 weeks, prematurity and low birth weight are the known risk factors of mortality. Therefore, pacemaker therapy should be considered in some cases after birth. However, there are only a few reports of pacemaker treatment for low birthweight infants with CCHB. Our report concerns a low birth infant with CCHB who underwent emergent pacemaker implantation in the first hour of life.

The 28-year old Polish Caucasian woman presented with Sjögren syndrome diagnosed 6 years earlier due to dry mouth and eyes, with Anti-SSA/Ro (151 RU/mL) and Anti-SSB/La (128 RU/mL) antiobdies, and was only on Plaquenil treatment. First trimester ultrasound screening showed normal anatomy and low risk of aneuploidy with a fetal heart rate (FHR) of 146 bpm (5th centile). Due to positive anti-Ro/La antibodies from the 16th week of pregnancy FHR was monitored weekly and at 18 weeks PR interval was 116 ms (Fig.1) whereas a week later a complete block was diagnosed by M-mode modality with an atrial rate of 134 bpm, a ventricular rate of 63 bpm (Fig. 2). Fetal echocardiogram demonstrated normal cardiac anatomy and CCHB with no signs of heart failure. After consultation with a prenatal cardiologist, dexamethason was prescribed. At 26 weeks of gestation, fibroelastosis (Fig. 3), cardiomegaly and a ventricular rate of 57 bpm were noticed, and salbutamol was introduced. After a week, the ventricular rate was 61 bpm. In the following weeks, a decrease in amniotic fluid, fetal growth retardation, and increased placental and uterine resistance were observed. At 36 weeks, because of anhydramnion, no weight gain, difficulties in fetal monitoring, decreased biophysical profile score to 7 and with cardiovascular profile of 7 (DV atrial reversal, holosystolic TR, heart size 0.45), it was decided to deliver the baby by cesarean section. The pediatric cardiovascular team set up an operating station in the delivery room. Physical examination after birth demonstrated a premature female infant in respiratory distress with a birth weight of 1600 g, Apgar scores 5/5/5, venous pH 7.198, and a ventricular rate of 45–50 bpm. The surfactant was administered, the child was intubated in moderate hypothermia (34.5 °C), and a limited median sternotomy was performed after umbilical arteriovenous vascular access. Two epicardial electrodes were fixed on the RV epicardial surface (apex and outflow tract) (Fig.4). Cardiac pacing with the external pacemaker was started at a rate of 110 bpm in VVI mode. After 2 months and the infant having reached 3.1 kg of body weight, the permanent pacemaker was implanted (Microny II SR+, St. Jude Medical) with stimulation in VVI mode. At the 2.5-year follow-up, the baby remains well without any complications.

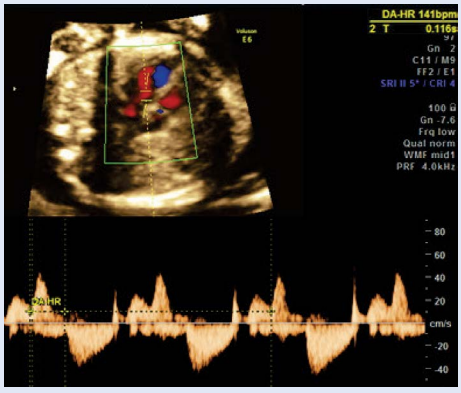

Figure 1. PR interval at 18weeks of gestation in a fetus with congenital complete heart block (CCHB)

To our knowledge, this is the first case of a pacemaker implantation due to autoimmune CCHB during the first hour of life, and with a long follow-up period, in Poland. This case report draws attention to the possibility of a sudden onset of the CCHB despite treatment with Plaquenil, difficulties in antenatal monitoring as Doppler parameters of umbilical artery, ductus venosus and extra-sinus slow FHR with no variation during fetal movements and uterine contractions

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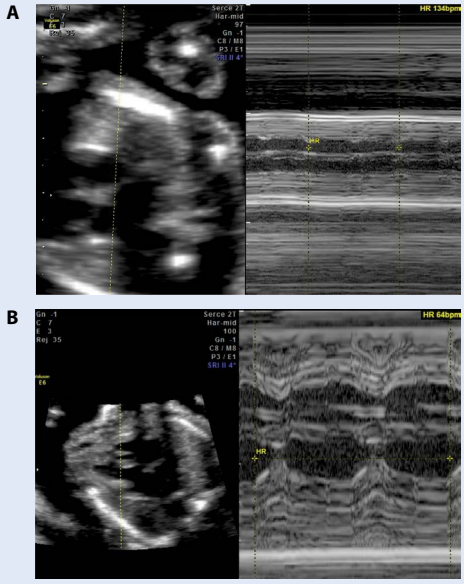

Figure 2. Atrial rate (**A**) and ventricular rate (**B**) at 19 weeks of gestation in a fetus with congenital complete heart block (CCHB)

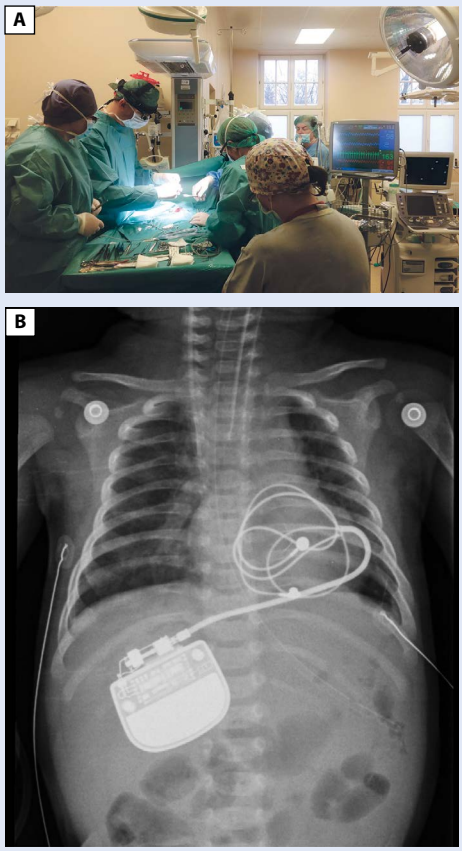

Figure 4. Pacemaker implantation in the delivery room in a newborn with congenital complete heart block (CCHB); **A.** Pediatric cardiac surgery team; **B.** Chest x-ray image of pacemaker implant

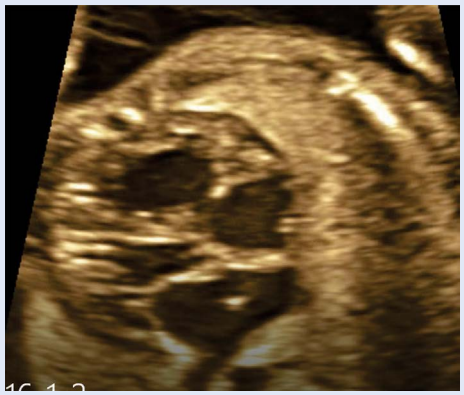

Figure 3. Four chamber view with endocardial fibroelastosis in a fetus with congenital complete heart block (CCHB)

are not useful. However planned delivery with a multidisciplinary team, followed by early pacemaker implantation may be an option for a severly affected newborns with CCHB. Temporary wires and external stimulation preceding the implantation of the permanent pacemaker seems to be the reasonable choice in newborns with low body weight.

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