# Successful Perinatal Management of a Very Low Birthweight Infant with Congenital Complete Atrioventricular Block

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## ABSTRACT

We report a very low birthweight infant with congenital complete atrioventricular block (CCAVB) who underwent delayed implantation of a permanent pacemaker without temporary pacing. The female infant was born at 30 weeks gestation and weighed 1422 g. After birth, the infant showed respiratory failure due to pleural effusion and respiratory distress syndrome at birth. The heart rate ranged between 50 and 55 bpm. The chest x-ray demonstrated dilated heart, but echocardiogram showed good systolic ventricular function. Respiratory failure was resolved after supportive treatment without temporary pacing. Mild heart failure due to low heart rate persisted, but was successfully managed by conventional heart failure therapy combined with nasal continuous positive airway pressure. She achieved a body weight gain to 1856 g at the age of 49 days and underwent implantation of a permanent pacemaker. We conclude that it is important to determine the timing of both delivery and pacemaker implantation for successful perinatal management of infants with CCAVB.

**KEYWORDS:** Congenital complete atrioventricular block (CCAVB), very low birthweight (VLBW) infant, permanent pacemaker

Congenital complete atrioventricular block (CCAVB) is classified into two groups: one is associated with congenital cardiac disease and another is associated with maternal autoimmune diseases. The incidence of the latter has been reported to be approximately 1:15,000 to 20,000 live births. However, the actual incidence is considered to be higher because many cases of CCAVB result in fetal demise.[1] The perinatal management of CCAVB has not yet been established. Fetal treatment, such as maternal β-stimulant administration, has rarely been successful. Many patients with CCAVB develop fetal heart failure and hydrops fetalis resulting in premature delivery. Early pacemaker treatment is sometimes required in cases of severe CCAVB. However, there are only a few reports of a pacemaker treatment for very low birthweight (VLBW) infants with CCAVB. Management of CCAVB commonly includes drug treatment such as β-stimulant, temporary cardiac pacing, and permanent pacemaker implantation. Drug treatment, however, is not always successful and temporary pacing carries a high risk for infection, especially in premature infants. We report a VLBW infant with CCAVB who underwent delayed implantation of permanent pacemaker after adequate body weight gain, whereas nasal continuous positive airway pressure (CPAP) was used to treat heart failure.

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## CASE REPORT

A 30-year-old woman, gravida 2 para 1, was referred for a fetal echocardiogram at 27 weeks gestation after routine Doppler interrogation of the heart demonstrated fetal arrhythmia. Fetal echocardiogram demonstrated normal cardiac anatomy. Interrogation of the rhythm by M-mode of atrial and ventricular contraction demonstrated dissociation of atrioventricular conduction. The arterial rate was 125 bpm and the ventricular rate was 54 bpm. Maternal assessment demonstrated positive antinuclear antibody (1:80, speckled pattern) and anti-Sjoegren's syndrome–A (SS-A) antibody (more than 500 U/mL). Intravenous ritodrine HCl was started to increase fetal heart rate, but was not effective. Repeat fetal ultrasound assessment demonstrated that fetal cardiomegaly and pleural effusion gradually increased at 29 weeks gestation. The decision was made to deliver the baby by cesarean section at 30 0/7 weeks gestation.

The weight was 1422 g and Apgar scores were 4 at 1 minute and 4 at 5 minutes. Physical examination demonstrated a premature infant in respiratory distress. There were no clinical signs of hydrops fetalis. An electrocardiogram showed that the rate of P wave was 125 bpm and the rate of QRS wave was 55 bpm. The blood pressure was 49/17 mm Hg. The chest x-ray demonstrated mild dilatation of the heart (cardiothoracic ratio, 58%), right pleural effusion, and lungs with neonatal respiratory distress syndrome (RDS; Fig. 1).

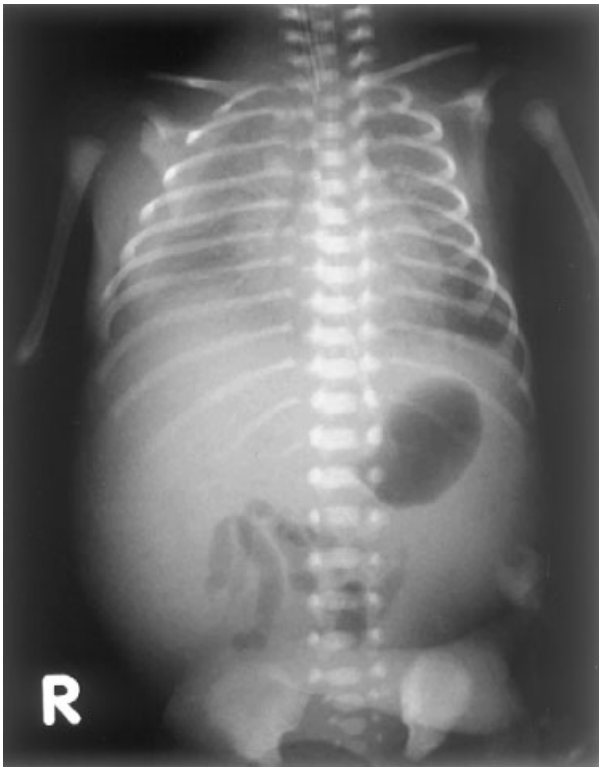


**Figure 1** Chest x-ray on admission. Note the right pleural effusion, lungs with respiratory distress syndrome, and cardiomegaly.

On echocardiogram, there was no congenital heart malformation, but dilatation of the heart and hyperdynamic cardiac function with fractional shortening of 59%. Surfactant replacement therapy for RDS was performed under mechanical ventilation. Dobutamine at 5 μg/kg/min and diuretics were used to support cardiac function against both low heart rate and pulmonary hypertension associated with RDS. The infant's cardiac function was also assessed by urine production, as well as echocardiogram. Urine output was sufficient under the supportive treatment and resulted in an adequate body weight loss of 12% at the age of 5 days. There was no need for temporary pacing. Both RDS and the pleural effusion were resolved by the age of 4 days.

After the resolution of pulmonary problems, the heart rate ranged between 45 and 50 bpm and the heart was still dilated. The infant was then managed to prevent heart failure and to promote adequate body weight gain. The diuretics (furosemide and spironolactone) combined with mild fluid restriction (120 to 140 mL/kg/day) was used to treat heart failure. In addition, nasal CPAP was used to prevent heart failure, as well as to reduce respiratory workload.

The infant gained body weight to 1856 g at the age of 49 days (37 weeks postconception), at which time she underwent implantation of a permanent pacemaker (MICRONY K SR model 2535K, St. Jude Medical, St. Paul, MN; VVI mode, stimulation rate 100 ppm, amplitude 3.0 V, width 0.31 milliseconds; Fig. 2). A unipolar epicardial lead was used, and the pulse generator, measuring 33 mm × 33 mm × 6 mm and weighing only 13 g, was implanted in a pocket made under the

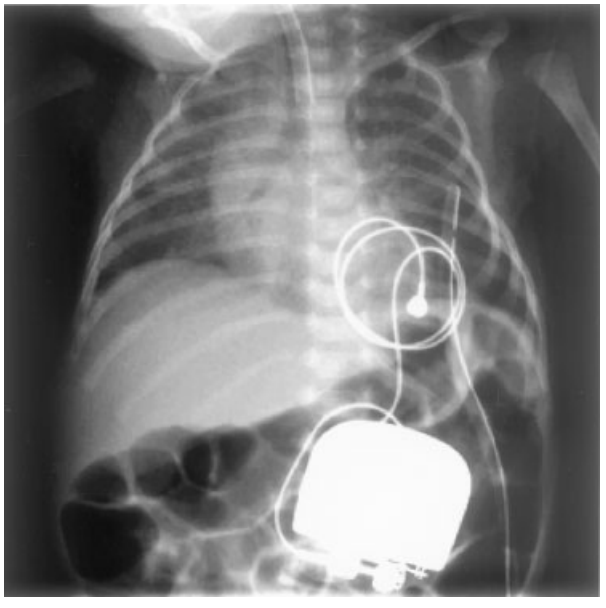

**Figure 2** Chest x-ray obtained at 49 days of life after implantation of permanent pacemaker. Note the location of the generator and the lead. Cardiomegaly has been resolved.

anterior rectus sheath. Surgery was performed without any complications. There was no respiratory problem associated with pacemaker implantation in the abdominal wall. She was discharged at the age of 71 days (40 weeks of gestation) with a weight of 2626 g. At the 1-year follow-up she remains well without any complications.

## DISCUSSION

Intrauterine diagnosis of CCAVB is based on complete dissociation between atrial and ventricular contraction on M-mode fetal echocardiogram. Between 30% and 37% of patients with CCAVB were diagnosed with an associated form of congenital heart disease.[2] In patients without cardiac anomalies, maternal connective tissue disease has been reported to play an important role in the development of CCAVB. Maternal anti-Ro (or related) antibodies from placental passage may injure fetal myocardium or conductive tissue.[3] The mother of the infant was positive for antinuclear antibody and anti-SS-A antibody, although she had no clinical symptoms of connective tissue disease.

In utero treatment of CCAVB remains controversial. Bierman et al[4] reported that maternal administration of corticosteroid was effective to reduce morbidity in fetuses with CCAVB. Buyon et al[5] suggested that plasmapheresis combined with corticosteroid therapy was effective in improving the outcome of infants with CCAVB. Furthermore, the administration of β-stimulant has been tried to increase fetal heart rate.[6]

The timing of delivery should be decided after careful evaluation of fetal cardiac function. Prompt delivery should be considered when signs of fetal heart failure or hydrops fetalis are observed. Donofrio et al reported that cardiovascular profile score combined with biophysical profile score was useful to determine the timing of delivery.[7] In this case, the decision was made to deliver because fetal heart failure was diagnosed based on progressive fetal cardiomegaly and fetal pleural effusion, and also because maternal administration of β-stimulant was not effective to increase the fetal heart rate.

Clinical features of neonates with CCAVB range from an absence of symptoms to severe heart failure. Heart rate is an important factor influencing cardiac function. A heart rate less than 60 bpm may cause heart failure in neonates. Administration of isoproterenol or atropine may be tried to increase the heart rate if the infant's heart rate is below 60 bpm, but these medications are rarely successful. Therefore, pacemaker therapy should be considered in many cases. Pacemaker implantation, however, is highly invasive in neonates, especially in VLBW infants. In addition, surgical procedures in premature infants carry a high risk of infection. The patient was a premature VLBW infant born at 30 weeks gestation and her condition was complicated by RDS. The heart rate ranged from 50 and 55 bpm, for which pacemaker implantation is indicated by American College of Cardiology/American Heart Association guidelines.[8] However, the patient showed a comparatively good cardiac function on echocardiogram and urine production was sufficient to reduce edema and pleural effusion. Therefore, we speculated that the timing of delivery was early enough to avoid the development of irreversible heart failure in this case.

The heart rate remained below 55 bpm and mild cardiomegaly persisted after the early neonatal period. To prevent heart failure, the patient was treated with mild fluid restriction, administration of diuretics, and the use of nasal CPAP. Nasal CPAP has been reported to be effective for the treatment of heart failure in adults.[9] Nasal CPAP is also used to treat apnea of prematurity.[10] Furthermore, we expected that the use of nasal CPAP could reduce respiratory workload in premature infants. As a result of these treatments, the patient showed an appropriate body weight gain without symptoms of heart failure.

A permanent pacemaker was implanted at the age of 49 days with body weight of 1856 g. A unipolar epicardial lead was used and the generator was implanted in a pocket made under the anterior rectus sheath to avoid erosion and infection due to very thin subcutaneous tissue in premature infants. Recent advances in pacemaker technology have made the pacemaker instrument smaller and lighter. There were no complications observed after implantation. A few case reports demonstrated that a permanent pacemaker was successfully implanted immediately after birth in VLBW infants with CCAVB.[7,11] In this case report, we describe a VLBW infant with CCAVB who safely underwent delayed pacemaker implantation.

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