Case Report

Successful Pacemaker Implantation for Congenital Complete Heart Block With Pulmonary Valve Stenosis in an Extremely Low Birth Weight Infant

Akio Kato, MD[1)], Masataka Kitano, MD[1)], Atsuya Shimabukuro, MD[1)], Seiichi Sato, MD, PhD[1)], and Masahiko Nishioka, MD[2)]

[1)] Department of Pediatric Cardiology, Okinawa Prefectural Nanbu Medical Center & Children's Medical Center, Okinawa, Japan
[2)] Department of Pediatric Cardial Surgery, Okinawa Prefectural Nanbu Medical Center & Children's Medical Center, Okinawa, Japan

Congenital complete heart block (CCHB) occurs because of transplacental antibodies such as anti-SS-A and anti-SS-B. CCHB is a potentially fatal condition, in particular in very/extremely low birth weight infants. Coexisting structural heart disease is another significant risk factor for CCHB. The survival rates after pacemaker implantation, either temporary or permanent, remain low. To the best of our knowledge, no criteria have been established for the minimum weight and earliest gestational age for treatment thus far. Herein, we present a case of an extremely low birth weight infant (ELBWI) with CCHB caused by positive anti-SS-A/Ro antibodies of the mother and significant pulmonary valve stenosis. The infant underwent temporary pacing implantation on the day of birth with a weight of 850 g. Percutaneous balloon pulmonary valvuloplasty was performed at the age of 106 days when body weight 2.6 kg. This consecutive approach provided a favorable outcome. We propose that epicardial temporary pacing could avoid postnatal heart failure in ELBWI with CCHB even when structural heart disease was present.

Keywords: congenital complete heart block, extremely low birth weight infant, structural heart disease, pulmonary valve stenosis, catheter intervention

Introduction

Congenital complete heart block (CCHB) occurs because of transplacental antibodies such as anti-SS-A and anti-SS-B antibodies, which are present in the mother's body. CCHB incidence ranges from 1 in 15,000 to 1 in 20,000 births.[1, 2)] Structural heart disease and low birth weight are recognized risk factors of CCHB.[3)] Herein, we report a case of an extremely low birth weight infant (ELBWI) with CCHB and severe pulmonary valve stenosis (PVS) who underwent pacemaker implantation and percutaneous balloon pulmonary valvuloplasty (BPV).

Case Report

A 35-year-old pregnant woman with elevated levels of anti-SS-A/Ro antibodies (>1200 U/mL; normal value, <10 U/mL) was referred to our hospital at 14 weeks of gestation. At 16 weeks of gestation, the fetus was diagnosed as second-degree atrioventricular heart block, with atrial and ventricular rates of 130 and 65 beats/minute, respectively. Since concurrent myocarditis was detected in the fetus, enteral dexamethasone therapy (4 mg daily) was initiated to prevent progression of fetal myocarditis. Unfortunately, atrioventricular conduction deteriorated into complete heart block at 19 weeks of

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Corresponding author: Akio Kato, MD, Department of Pediatric Cardiology, Okinawa Prefectural Nanbu Medical Center & Children's Medical Center, 118–1 Arakawa Haebaru, Okinawa 901–1105, Japan
E-mail: akki.kato@gmail.com
Akio Kato (https://orcid.org/0000-0002-8727-5634)
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gestation. Eventually, the mother was hospitalized for meticulous monitoring and ritodrine was commenced at 26 weeks' gestation. At that time, the fetus had a cardiovascular profile score (CVPS) of 6; due to pericardial effusion, a cardiothoracic area ratio (CTAR) 45%, and pulsatile umbilical venous blood flow. We detected hyperintensity of the pulmonary valve, but no acceleration of blood flow across it. The fetal heart rate slightly increased from 51 to 56 beats/minute.

At 27 weeks of gestation, ritodrine had to be discontinued because of maternal liver dysfunction. Fetal ascites appeared at 27 weeks + 2 weeks. At 27 + 3 weeks, fetal ascites increased although CVPS was 5 and biophysical profile score was 8, CTAR got worse to 55%, fetal movement decreased, and the heart rate dropped to 48 beats/minute; these changes necessitating an urgent cesarean section. The infant born was a female weighing 850 g. Apgar scores were 4 and 5 at 1 and 5 minutes, respectively. Her heart rate was 48 beats/minute. Immediately after birth, the infant was intubated and transferred to the operating room. Pulmonary surfactant was administrated into the trachea and umbilical arterial and venous catheters were placed.

Subsequently, a temporary pacing lead was implanted as follows. Through a median sternotomy, an Osypka heart wire (TME 56-T) bipolar (Osypka, Tokyo, Japan) measuring 60 cm in length was affixed to the anterior epicardium of the right ventricle using 7-0 monofilament sutures. Then, pacing was started at 80 beats/minute. Postoperative echocardiography illustrated that the diastolic left ventricular intradiameter was 10.2 mm and fractional shortening was 30%. It also showed myocardial hypertrophy, for which hydrocortisone (3 mg/kg/day) was initiated in order to prevent further myocardial damage and late circulatory dysfunction in the infant. This treatment was tapered off over a period of 3 weeks. PVS was another lesion detected. The pacemaker lead was replaced at 38, 73, and 124 days of age because pacing capture threshold was elevated beyond an acceptable level.

Regarding PVS, the annular diameter of the pulmonary valve was 4.4 mm (Z-score −1.6) at birth, and blood flow velocity was 3.2 m/s across the orifice. The velocity increased up to 4.5 m/s after the third placement of the pacemaker lead. Right ventricular pressure was estimated higher than left ventricular pressure. The foramen ovale shunted right-to-left, leading to persistent hypoxemia. Percutaneous BPV was performed at 106 days of age (2.6 kg body weight) since oxygen saturation fell below 80% during crying.

Finally, at 4 months of age (weight, 3.6 kg), an intraperitoneal generator (ZYPHYR SR 5620, Abbott Medical Japan LLC., Tokyo, Japan) and an epicardial ventricular lead (CapSure Epi model 4965, Medtronic INC., Minneapolis, Minnesota, USA) were implanted onto the inferior surface of the right ventricle.

The infant was discharged 19 days after implanting the permanent pacemaker. At 15 months of age, a generator replacement was carried out because the skin became rather thin around the generator pocket.

Discussion

Treatments for CCHB have improved. Still, high-risk cases do not diminish markedly. Recent reports have identified several risk factors leading to prenatal or postnatal death in infants with neonatal cardiac lupus and in fetuses with second- or third-degree atrioventricular block. These include the maternal/intrauterine diagnosis of myocarditis, endocardial fibroelastosis, systemic lupus erythematosus, Sjögren's syndrome, AV block diagnosis before 20 weeks, ventricular rate of < 50 beats/minute, fetal hydrops, comorbid congenital heart

Table 1 Prior reports of pacemaker therapy at birth for congenital complete heart block in patients born with 28 weeks of gestation or less

Year	Gestational week (weeks)	Birth weight (g)	Heart rate at birth (/min)	Age at operation for initial pacing (days)	Pacemaker type	Structural heart disease	Outcome
2007[7]	27	980	55	2	Epicardial	None	alive
2000[6]	26	830	50	0	Transthoracic	None	dead
2012[8]	28	650	—	1	permanent	None	dead
	28	840	—	1			alive
Our case	27	850	48	0	Epicardial	PVS	alive

PVS: pulmonary valve stenosis

disease, and presence of left ventricular dysfunction at diagnosis.[3–5] Our patient exhibited quite high risks for CCHB, which was diagnosed at 19 weeks of gestational age, extremely early term and low birth weight of 850 g, and significant pulmonary valve stenosis. After extensive discussions among the medical staff as well as with the patient's family, an approach for aggressive treatment was selected, and the approach turned out to provide a favorable outcome.

CCHB treatment is particularly challenging in ELBWIs; only a few cases having been reported in literature. In previous reports, pacemaker implantation was performed in four ELBWIs before 28 weeks of gestational age (Table 1).[6–8] Among the four cases, only two survived, with the earliest gestational age and minimum weight being 27 weeks and 840 g, respectively. To the best of our knowledge, this case report represents the successful implementation of pacemaker implantation at the lowest weight of 27 weeks gestation.

The approach how to implant a pacemaker for CCHB remains undetermined in ELBWI. Nürnberg et al employed transthoracic pacing in a 26-week-old infant weighing 830 g,[6] whereas Shepard et al performed permanent pacemaker implantation in infants weighing 630 g and 840 g.[8] There was only one case report describing use of temporary pacing in ELBWI.[7] For premature infants, we considered that temporary pacing via an epicardial approach was reasonable, given their small thorax and abdomen as well as their immature skin.

The prognosis is extremely poor in low birth weight infants with CCHB and coexisting structural heart disease.[5, 9] Previous reports indicated that structural heart disease occurred in 53% of CCHB cases, with a mortality rate exceeding 45%.[5] The survival rate was extremely low in those with heterotaxy.[5, 9] In the present patient, PVS was considered moderate or severer. The obstruction got worse postnatally causing hypoxia. Percutaneous BPV, which is less invasive than surgery, was safely achieved. We attributed the favorable outcome to the fact that surgery was not mandatory for the degree of PVS; the lesion was responsible for hypoxic circumstance but did not pose a loss of cardiac output.

Conclusion

The present case demonstrates that preterm ELBWIs with CCHB and coexisting severe PVS were successfully treated with combination of pacemaker implantation and percutaneous BPV. Gestational age of over 27 weeks and birth weight exceeding 800 g may be a potential indicator for survival with appropriate therapy. This report, although represents a single patient, implies that these cutoff figures would be reasonable and that the prognosis would also depend on how severe the coexisting structural heart diseases were.

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Author Contribution

KA drafted the original manuscript. KM and SA had revised this manuscript, and SS and NM had finally reviewed the manuscript and approved the published version.

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