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Permanent Pacemaker Implantation in Neonates with Congenital Complete Atrioventricular Block: A Case Series

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Abstract

Introduction Congenital complete atrioventricular (AV) block manifests in approximately 1 in 20,000 live births and poses significant risks of mortality and morbidity both prenatally and postnatally. Its etiology can stem from either immune or non-immune mediated pathways. Immune-mediated AV block is a complex condition often linked to the transplacental transmission of maternal autoantibodies, particularly anti-Ro/SSA and/or anti-La/SSB antibodies. Implanting permanent pacemakers in neonates with congenital complete AV block presents technical challenges due to their small size, the presence of concurrent structural heart anomalies, and the rapid growth rate typical of infants. Case description We encountered five instances of congenital complete atrioventricular (AV) block in neonates exhibiting persistent low heart rate post-birth. Among these cases, two patients had mothers with a history of autoimmune disease, specifically Sjogren's syndrome with positive Ro antibodies and receiving methylprednisolone, while the remaining three cases had no history of autoimmune disease. Additionally, one case presented with a congenital anomaly, namely anorectal malformation, and three cases were complicated by sepsis during hospitalization. Diagnosis was established perinatally, with intervention involving immediate permanent pacemaker implantation after birth in two cases, and in the remaining three cases, permanent pacemaker implantation occurred within the subsequent days. Encouragingly, all five cases exhibited favorable outcomes without complications. Conclusion Congenital complete AV block represents a rare yet critical condition necessitating prompt recognition. Early diagnosis and treatment, particularly through permanent pacemaker implantation, play pivotal roles in mitigating morbidity and mortality rates associated with this condition, ultimately leading to improved patient outcomes.

Keywords: *Permanent pacemaker installation, atrioventricular, neonates*

Introduction

Cardiac conduction disorders are infrequent conditions found in newborns and youngsters. They can manifest in hearts that are structurally sound or in conjunction with congenital heart disease (CHD), stemming from various causes (Baruteau et al., 2016) . Congenital complete heart block is characterized by a total blockage of the atrioventricular pathway either before birth, at birth, or within the initial month after birth. Its occurrence is estimated to be around one in every 15,000 to 20,000 live births according to available literature(Bora et al., 2022).

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Congenital complete heart block (CCHB) is associated with a significant mortality rate, and typically, neonatal cases of this condition are irreversible (Ju et al., 2021).

Congenital atrioventricular (AV) block is a rare but important cause of neonatal bradycardia (Mitra et al., 2013). Congenital AV block can be passively acquired via an autoimmune process affecting the developing heart due to the transplacental passage of maternal anti-Ro/SSA and/or anti-La/SSB autoantibodies (Baruteau et al., 2016). Pathophysiology involves local inflammation, calcification, and fibrosis of the cardiac conduction system. CCHB is also associated with CHD in 14–42% of cases, such as left atrial isomerism (Hernstadt et al., 2022).

Individuals with complete congenital heart block may experience diminished cardiac function or reduced cardiac output. Administering early temporary pacing serves as a transitional measure to permanent pacemaker implantation in high-risk patients, mitigating the detrimental effects of severe bradycardia and even asystole. According to the AHA 2012 guidelines, permanent pacemaker placement was recommended for symptomatic infants with complete congenital heart block, cases of nonreversible AV nodal disease, and asymptomatic infants exhibiting a ventricular rate below 55 bpm, or below 70 bpm in instances of complete congenital heart block accompanied by significant cardiac structural abnormalities (Ju et al., 2021).

Despite advances in antenatal and neonatal care, early morbidity and mortality of CCHB are high. Risk factors for poor outcome include fetal hydrops, low ventricular rate (heart rate (HR) < 55 beats per minute (bpm) at presentation or one which drops < 50 bpm), and associated CHD. Prematurity is a risk factor for death, though data on neonatal morbidity due to CCHB are scarce (Hernstadt et al., 2022). We hereby report a case series of congenital AV block in neonates' patients in NICU using the permanent pacemaker implant (SpheraT™ SR MRI, SURESCAN™ model SPSR01-Medtronic).

Case Description

Case 1

Baby Y, a female neonate delivered via caesarean section at a gestational age of 38 weeks due to fetal bradycardia and total AV block detected by ultrasonography performed at the onset of labor. The decision for caesarean section was prompted by the observation of fetal bradycardia using a fetoscope in the hospital setting. The mother had a medical history of Sjogren's syndrome with positive ro antibodies and was undergoing treatment with methylprednisolone. The newborn exhibited spontaneous respiration upon delivery but maintained a persistently low heart rate of 35-45 bpm thereafter. Laboratory investigations revealed neutropenia, thrombocytopenia, elevated levels of C-reactive protein (CRP), and procalcitonin. Chest X-ray demonstrated cardiomegaly, while ECG confirmed the presence of total AV block. Echocardiography revealed dilated chambers, an atrial septal defect (ASD), patent ductus arteriosus (PDA), with an ejection fraction of 68%. Management included fluid and nutritional therapy, antibiotics for sepsis, dopamine, hydrochlorothiazide, spironolactone, and plans for permanent pacemaker implantation. Following resolution of sepsis, the permanent pacemaker was successfully implanted, resulting in an improved heart rate of 110-120 bpm. The patient was discharged after 12 days without complications.

Case 2

Baby DAD, a female neonate delivered via caesarean section at a gestational age of 38 weeks due to fetal bradycardia and tricuspid valve dilatation diagnosed by ultrasonography conducted

at the onset of labor. The mother had a medical history of Sjogren's syndrome with positive ro antibodies and was receiving methylprednisolone treatment. The newborn exhibited spontaneous respiration upon delivery but maintained a persistently low heart rate of 40-50 bpm thereafter. Laboratory findings were within normal limits. Immediate surgical intervention was undertaken to implant a permanent pacemaker shortly after birth. Subsequent echocardiography following the pacemaker installation revealed dilation of both the right and left ventricles, along with an atrial septal defect (ASD) and an ejection fraction of 54%. Further management included fluid, nutritional therapy, and diuretics. Post pacemaker implantation, the patient's condition improved with a heart rate of 120-130 bpm, and she was discharged after 10 days without any complications.

Case 3

NA, a female neonate delivered via caesarean section at a gestational age of 38 weeks due to fetal bradycardia and total AV block diagnosed by ultrasonography performed at the onset of labor. The mother had no history of lupus or Sjogren's syndrome. The newborn exhibited spontaneous respiration upon delivery but maintained a persistently low heart rate of 50-55 bpm thereafter. Laboratory findings were within normal limits. Immediate surgical intervention was undertaken to implant a permanent pacemaker shortly after birth. Subsequent echocardiography following the pacemaker installation revealed a small patent ductus arteriosus (PDA) with an ejection fraction of 69%. Further management included fluid and nutritional therapy. Post pacemaker implantation, the patient's condition improved with a heart rate of 100-120 bpm, and she was discharged after 23 days without any complications.

Case 4

Baby FAR, a female neonate who was delivered through caesarean section at gestational age of 38 weeks. The indication for caesarean section was fetal bradycardia diagnosed by ultrasonography done at the onset of labour. Mother no history of lupus or Sjogren's. Baby was delivered with spontaneous respiration and no defecation from birth. Thereafter, the baby remained clinically stable apart from persisting low heart rate of 60-70 bpm. The patient has an anorectal malformation. Laboratory findings with elevation CRP, thrombocytopenia, and anemia. Echocardiography revealed persistent foramen ovale with left to right shunt. Management includes the installation of a permanent pacemaker and correction of anorectal malformation accompanied by a colostomy on day 6 and other therapies consisting of fluid, nutritional therapy, diuretics and antibiotic. Echocardiography post installation of permanent pacemaker revealed no persistent foramen ovale. After an installation of a permanent pacemaker, the patient's condition improved heart rate of 100-130 bpm and the patient was discharged after 18 days and with no complications.

Case 5

SN, a male neonate delivered via caesarean section at 38 weeks gestation due to fetal bradycardia and total AV block detected by ultrasonography at the onset of labor. The mother had no history of lupus or Sjogren's syndrome. The infant demonstrated spontaneous respiration upon delivery but persisted with a low heart rate ranging between 50-60 bpm thereafter. Laboratory investigations indicated thrombocytopenia and elevated C- reactive protein (CRP) levels. Echocardiography revealed the presence of a small patent ductus arteriosus (PDA) and an atrial septal defect (ASD) secundum.

Management involved the immediate placement of a permanent pacemaker on the first day of life, alongside other therapeutic interventions including fluid administration, nutritional therapy, and antibiotics for suspected sepsis. Subsequent echocardiography post pacemaker implantation demonstrated a small ASD secundum remaining.

Following the pacemaker procedure, the patient's clinical condition exhibited improvement, with a heart rate rising to 100-120 bpm. After 17 days of comprehensive care, characterized by attentive monitoring and therapeutic measures, the patient was discharged without experiencing any complications.

Table 1: Demographics, Clinical and Laboratory Features, and Outcome of the Three Cases.

Parameter	Case 1	Case 2	Case 3
Sex	Female	Female	Female
Symptom	-	-	
Diagnosis	Total AV Block Sepsis, ASD and PDA	Total AV Block and ASD	Total AV Block and PDA
Mothers' disease	Sjogren syndrome with ro positive	Sjogren syndrome with ro positive	-
Blood pressure (mmhg)	83/37	84/46	54/39
Heart rate (beats/min)	35-45	40-50	50-55
Respiratory rate (times/min)	45-60	45-55	40-55
Temperature instability	Yes	No	No
Oxygen saturation (%)	92-98	91-95	92-97
White blood count ($10^3/\mu L$)	15.27	13.28	18.41
Hemoglobin (mg/dL)	16.4	14.9	12.6
Platelets ($10^3/\mu L$)	157	122	551
CRP (mg/dl)	10.8	3.1	16
Chest X-Ray	Cardiomegaly	Normal	Normal
ECG	Total AV Block	-	-
Echocardiography	Dilated chamber, Small secundum ASD, Tiny PDA, EF 68%,	Dilated right and left ventricles and ASD with an ejection fraction of 54%	Total AV Block, small PDA with an ejection fraction of 69%.
Temporary pace maker (TPM)	-	-	-
Permanent pace maker (PPM)	12nd days	Immediately after birth	Immediately after birth
Heart rate (beats/min) after PPM installation	115-125	120-130	100-120
Outcome	Discharged	Discharged	Discharged

Table 1: Demographics, Clinical and Laboratory Features, and Outcome of the Two Cases (Cont').

Parameter	Case 4	Case 5
Sex	Female	Male
Symptom	No defecation from birth	
Diagnosis	Total AV Block, PFO, sepsis and anorectal malformation	Total AV Block, PDA, ASD and sepsis
Mothers' disease	-	-
Blood pressure (mmhg)	87/51	91/65
Heart rate (beats/min)	60-70	50-60
Respiratory rate (times/min)	45-55	40-55
Temperature instability	No	Yes
Oxygen saturation (%)	92-98	92-97
White blood count ($10^3/\mu L$)	21.54	47.9
Hemoglobin (mg/dL)	9.8	17
Platelets ($10^3/\mu L$)	134	35
CRP (mg/dl)	76.2	123.8
Chest X-Ray	Normal	Normal
ECG	Total AV Block	
Echocardiography	Persistent foramen ovale	Small PDA and ASD secundum
Temporary pace maker (TPM)	-	-
Permanent pace maker (PPM)	6th days	1st day
Heart rate (beats/min) after PPM	100-130	100-120
Outcome	Discharged	Discharged

Discussion

Congenital heart block (CHB) is an uncommon condition, with elevated mortality rates when occurring alongside structural congenital heart abnormalities. This neonatal ailment is infrequent, presenting in approximately 1 in 20,000 live births. Detection may occur prenatally, often manifesting as either first- or second-degree AV block, though the majority of cases exhibit a critical third-degree, complete AV block which can be fatal. In certain instances, there may also be a dangerous co-occurrence with cardiomyopathy (Di Mauro et al., 2013). AV block refers to a disruption in cardiac conduction characterized by irregular propagation of the electrical impulse, stemming from structural or functional irregularities. When identified during prenatal screenings, at birth, or within the initial month of life, it is classified as congenital. In this study, we present seven case reports of neonatal patients diagnosed with congenital AV block at birth, all of whom underwent management through permanent pacemaker intervention.

The etiology of congenital atrioventricular block (CAVB) encompasses various factors, with a significant portion stemming from structural congenital heart conditions such as corrected transposition of the great artery and left atrial isomerism, a form of heterotaxy syndrome, constituting over half of the cases. These structural abnormalities are considered non-immunological in nature. However, another causative mechanism involves immunological processes, wherein maternal autoantibodies traverse the placenta into the fetal circulation. Notably, the autoantibodies implicated in CAVB are anti-Ro/Sjögren's syndrome antigen A (SSA) and anti-La/Sjögren's syndrome antigen B (SSB) (Hansahiranwadee, 2020). Fetal tissue injury is believed to be influenced by the transplacental passage of maternal IgG autoantibodies, primarily mediated by FcγR receptors. Specifically, anti-SSA/Ro and anti-SSB/La antibodies attach to fetal cardiomyocytes, impeding the natural elimination of apoptotic cells, thereby triggering an inflammatory response and fibrosis within the cardiac conduction system. Additionally, potential mechanisms include the cross-reactivity of SSA/SSB antibodies and the modulation or inhibition of L-type Calcium channels by these autoantibodies. Some researchers have explored the electrophysiological and molecular mechanisms underlying congenital heart block, suggesting that anti-SSA/Ro antibodies may directly induce arrhythmogenic activity. Ultimately, these processes culminate in myocarditis, hemorrhage, fibrosis, calcification, and necrosis within the conduction system, leading to the development of varying degrees of heart block, myocardial dysfunction, and/or endocardial fibroelastosis (Yildirim et al., 2013) Clinically, the condition may present with bradycardia, characterized by heart rates below 100 beats per minute (bpm), along with pericardial effusion, ventricular dilation, increased echogenicity of the atrial walls due to fibrosis, and decreased ventricular contractility. Echocardiogram findings may reveal signs of inflammation and likely myocarditis, endocardial fibroelastosis, as well as manifestations such as ascites and fetal hydrops. A ventricular response of less than 55 bpm, prematurity, and the presence of complex congenital defects serve as indicators of a poor prognosis, correlating with higher mortality rates (Melim et al., 2022).

A thorough medical history, particularly of the mother, is essential to obtain an accurate and timely diagnosis. In high-risk pregnancies, fetal echocardiographic cardiac monitoring should begin in the 16th week and continue weekly until the 24th week, and thereafter fortnightly until birth (Melim et al., 2022). The diagnosis of congenital heart block (CHB) detected prenatally is commonly achieved through either clinical auscultation or routine obstetrical ultrasound, with confirmation typically conducted via fetal echocardiogram employing Doppler techniques.

This diagnostic procedure serves the dual purpose of identifying the level of block and excluding major concurrent structural heart defects, such as left atrial isomerism with or without atrioventricular septal defects, and ventricular inversion. These structural abnormalities are linked to heart block but are not antibody related. Furthermore, the fetal echocardiogram can assess for associated myocarditis by observing decreased contractility and secondary indicators like cardiomegaly, tricuspid insufficiency, pericardial effusion, or the onset of hydrops fetalis (Nkoke, 2016). In our case, the diagnosis may have been made in-utero like that reported by Dey et al. and might have dictated the attitude with respect to management of the pregnancy with serial echocardiograms and treatment after delivery.

Patients with complete congenital heart block may exhibit compromised cardiac function or reduced cardiac output. In cases of severe bradycardia, treatment options include intravenous administration of isoproterenol, atropine, epinephrine, and dopamine. However, it's important to note that no medication has been definitively shown to enhance chronic sinus and atrioventricular nodal function beyond acute management (Ikuta et al., 2017). Early temporary pacing serves as a bridging strategy to permanent pacemaker implantation in high-risk patients, mitigating the adverse effects of severe bradycardia or even asystole. Intravenous administration of isoproterenol, atropine, epinephrine, and dopamine has also been documented in clinical practice. The decision to implant a permanent pacemaker is primarily guided by the presence of symptomatic bradycardia and the severity of the AV block. According to existing guidelines and clinical practice, indications for pacemaker implantation include symptomatic bradycardia, significant AV block with compromised cardiac function, or evidence of hemodynamic instability. The threshold heart rate for intervention varies but commonly includes ventricular rates below 55 bpm or less than 70 bpm in the presence of major structural heart defects. Pacemaker implantation is performed in approximately 12–53% of neonates with complete congenital heart block, and a majority require long-term pacing. Complications following pacemaker implantation, occurring in up to 25% of cases, may include infections, cardiac perforations, thromboembolisms, lead fractures, and, over the long term, pacemaker-induced myocardial dysfunction (Ju et al., 2021). In our case, the patient was clinically symptomatic with bradycardia after birth. We opted against temporary or permanent pacemaker insertion and continued to monitor the patient. The clinical course spontaneously improved, and the patient was discharged with permanent pacemaker implantation.

Complex CHB has a higher morbidity and mortality than isolated CHB and this is determined more by the underlying structural congenital heart disease than by the need for a pacemaker. Mortality is highest in the neonatal period and association with major cardiac malformations is a bad prognostic sign. The outcome for patients with congenital heart block depends largely on the presence or absence of underlying structural heart disease, a heart rate less than a critical value, frequently quoted as 55 bpm and the presence or absence of congestive heart failure. If the heart block is diagnosed as a bradycardia during the fetal period, there is a very high rate of fetal and neonatal loss. There is a higher association of congenital heart block occurring with congestive heart failure in utero, and thus a poorer prognosis (Chukwuebuka et al., n.d.). In this patient, permanent pacemaker implantation was carried out early so as to prevent complications such as congestive heart failure so that the patient's outcome was very good.

The ultimate goal of permanent pacemaker implantation in neonates with congenital complete AV block is to optimize cardiac function, alleviate symptoms, and improve long-term prognosis. In our case series, we observed favorable outcomes following pacemaker implantation, including resolution of bradycardia, restoration of adequate heart rate, and

improvement in overall clinical status (Chukwuebuka et al., n.d.). However, it is essential to recognize that outcomes may vary among individuals and are influenced by factors such as the underlying etiology of AV block, associated cardiac abnormalities, timing of intervention, and postoperative management (Chukwuebuka et al., n.d.). Long-term follow-up studies are warranted to assess the durability of pacemaker function, potential complications, and the impact on neurodevelopmental outcomes in affected neonates.

In Indonesia, the trend of Permanent Pacemaker Implantation (PPI) in neonates can be compared to other countries based on available data. While specific data in Indonesia may not be directly available in the provided references, trends from other countries can provide insights. For instance, a study by Misumida et al. (2018) highlighted a trend towards earlier PPI in the United States, with a median of 1 day in 2014 compared to previous years (Misumida et an., n.d.). This indicates a potential shift towards more prompt interventions. Similarly, research by Bhat et al. (2018) suggested that in India, PPI is often done for survival rather than just quality of life improvement, which could reflect trends in some developing countries like Indonesia (Bhat et al., n.d.).

Conclusion

We documented five instances of bradycardia associated with AV blocks. The presence of predisposing factors such as a prior autoimmune disorder history may suggest a likelihood of AV block development in infants. Timely identification followed by decisive intervention involving permanent pacemaker implantation and concurrent management of comorbid conditions yields a favorable prognosis.

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