CASE REPORT

Placement of permanent pacemaker in a low-birth-weight infant with congenital heart block: a case report

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Abstract

One of the rare diseases with a high mortality rate in infants is congenital heart block (CHB) with neonatal lupus erythematosus (NLE) as the most common cause. A permanent pacemaker (PPM) is indicated for symptomatic bradycardia. The choice of PPM in the paediatric population is different from that in the adult population because of several reasons like small size, account of somatic growth, and difference in physiological changes. Here, we present a case in which a 2.6 kg and 45 days old baby with CHB secondary to NLE was successfully treated with a single-chambered adult-sized PPM with epicardial lead. According to our knowledge, this is the smallest baby in Pakistan in which PPM has been implanted.

Keywords: Neonatal lupus erythematous, infant low birth weight, cardiac pacemaker, permanent pacemaker implantation (ppm), congenital complete heart block.

DOI: 10.47391/JPMA.6674

Submission completion date: 13-04-2022
Acceptance date: 13-10-2022

Introduction

Congenital heart block (CHB) is a rare but potentially life-threatening condition with an estimated incidence of 1 in 25000 infants.[1] It is defined as the presence of cardiac conduction abnormalities in utero or within the first few days of life. The majority (90%) of the CHBs are due to maternal autoimmune disease or congenital structural heart disease. The rest (10%) are due to idiopathic causes. Neonatal Lupus Erythematosus (NLE) is the most common cause of autoimmune CHBs, and it occurs as a result of transplacental passage of maternal anti-Ro/SSA or less commonly anti-SSB/La (anti-La) and anti-RNP autoantibodies to the foetus. The only effective treatment in infants in the placement of permanent pacemaker (PPM). It is indicated when there is heart rate of less than 55 beats/min.[2]

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While pacemakers are the mainstay of treatment in CHB infants, they comprise <1% of all pacemaker implantations. PPMs are classified into a single or dual-chamber device with epicardial or endocardial pacing. Over the years, the choice of pacemaker in infants has been a topic of debate. Various issues such as small body size, account of somatic growth, and associated pathophysiology in the paediatric population makes the choice of PPM distinctive from the adult population.[3] In developing countries, availability and cost play a major role in the selection of pacemakers due to the scarcity of resources.

Here, we present a case of a 45-day old baby who was diagnosed with CHB due to NLE. We placed a single-chambered adult size pacemaker. Treatment was successful with an uneventful recovery.

Case Report

A 35-year-old primigravida (G1 P1+0) conceived via in-vitro fertilization. She was advised foetal echo at 20 weeks of gestation, which showed foetal bradycardia with complete heart block. There were no associated antenatal comorbidities. During further antenatal visits, the diagnosis of lupus was suspected and anti-Ro/SSA and anti-SSB/La (anti-La) antibodies tests were done which came out to be positive. Treatment for mother's lupus was started and foetal monitoring was being done weekly.

Foetal heart rate was between 100 bpm to 110 bpm. At 30 weeks of gestation, the baby was delivered via C-Section. After 3 weeks in NICU, the baby was discharged on low dose oxygen therapy with continuous monitoring of heart rate. The weight at the time of discharge was 1.7 kg.

On 45th day of life, the baby's heart rate dropped from 130 bpm to 40 bpm. He was brought to the Emergency department at Liaquat National Hospital & Medical College/ Karachi in August 2020 where a 3rd-degree heart block was confirmed on the ECG (Figure 1). After counselling the parents, consent was obtained, and the patient was prepared for PPM placement.

Since the baby weighed only 2.6 kg, it was planned to use a single chamber PPM (Sensia SESR01, Medtronic inc.). Through a 5 cm midline incision over the xiphoid process,

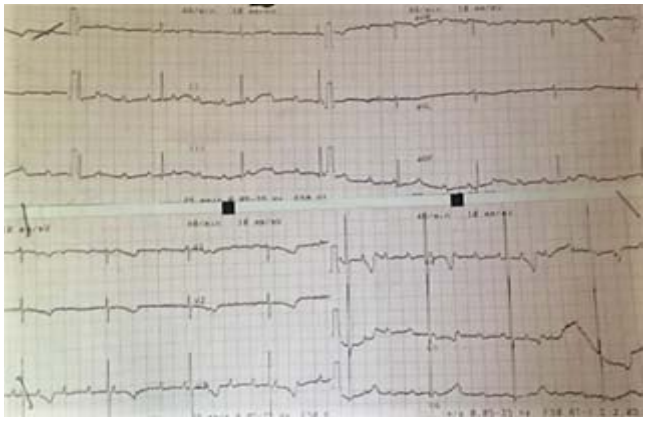

Figure-1: Twelve lead electrocardiogram (ECG) of baby showing complete heart block with no relation between P wave and QRS complex and junctional escape rhythm as 40 bpm

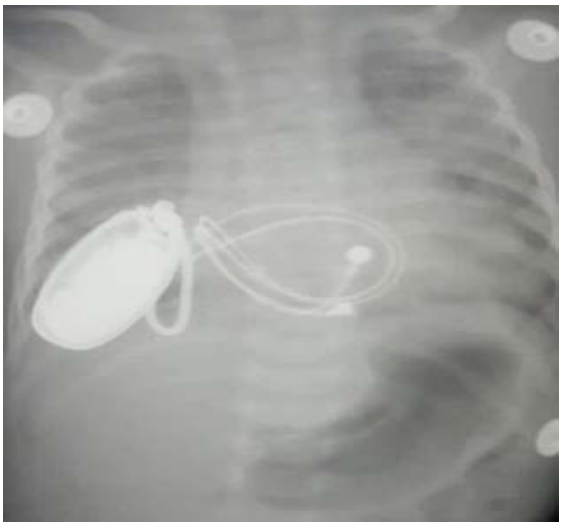

Figure-2: Post-operative X-ray showing PPM implantation with epicardial leads.

the pericardium was entered. The pacemaker wire leads were fixed to the epicardium of the right ventricle using 5-0 polypropylene sutures. The pacemaker box was placed in the right pleural cavity and secured by sutures to the diaphragm to prevent it from migrating. The electrodes were connected to the pacemaker generator, which was set at lower 90 bpm and upper 130 bpm. The sensitivity of ventricular lead was 2.8 mV. The wound was then closed in a routine fashion.

The postoperative recovery was uneventful, and the baby was discharged on the 3rd day and was advised for follow-ups. Chest X-ray (Figure 2) and PPM interrogation (Table) were done at the time of discharge. The patient visited at 1 year follow up in August 2021 where strength

Table: Permanent Pacemaker interrogation and results

Mode		**Rate Response**		**Battery status**	
Mode	VVIR (ventricular pacing, ventricular sensing, inhibition response and rate-adaptive)	Upper Sensor Rate Setpoint	40	Estimated remaining longevity	9.5 years. 8-11 years
Rates		Ventricular Lead		Voltage/Impedance	2.78 V/ 100 ohms
Lower Rate	90 ppm	Amplitude	3500 V	Lead Summary	Ventricular
Upper Sensor Rate	130 ppm	Pulse Width	0.40 ms	Programmed Output	3500 V/ 0.40 ms
ADL (Activities of daily living) Rate	95 ppm	Sensitivity	2.80 mV	Capture	Adaptive
Ventricular Refractory	330 ms	Sensing Assurance	On	Measured R wave programmed Sensitivity	2.80 mV
Rate Response		Pace Polarity	Bipolar	Measured Impedance	1393 ohms
Optimization	On	Lead Monitor	Monitor only	Lead Status	OK
ADL Response	3	Maximum Impedance	4000 ohms	Lead Model	496835
Exertion Response	3	Minimum Impedance	200 ohms	Implanted	24/08/20
ADLR Percent	2.0%	Motor Sensitivity	8		
Activity Threshold	Medium/Low	Capture Management	Adaptive		
Activity Acceleration	30 sec	Amplitude Margin	2x		
Activity Deceleration	Exercise	Min Adapted Frequency	Day at rest		
High-Rate Percent	0.2%	Acute Phase	112 days		
ADL Rate Setpoint	15	V sensing during search	Adaptive		

Figure-3: Strength amplitude test of ventricle.

duration amplitude test of ventricle was done (Figure 3) and amplitude was changed from 2.0 to 3.5 mV.

Parental consent was obtained for publishing the case report.

Discussion

The development of pacemakers has revolutionized the treatment of CHB. Since their discovery, innovations have been made in device technology, lead design, battery lifespan, and software algorithms. The choice of PPM in infants is still a dilemma as there is not enough evidence in literature in the form of RCTs. However, the consensus is to consider patient's size and anatomy.[4]

Generally, dual-chamber PPMs are preferred as they better synchronize the atrial and ventricular rhythm reducing the chances of pacemaker syndrome; a myriad of signs and symptoms of heart failure and hypotension after placement of PPM due to loss of atrioventricular synchrony.[5] Dual-chamber PPM also decreases the risk of other complications such as atrial fibrillation, stroke, and death.[6] However, they are not recommended for small infants because of their size. However, the smaller size of generator in single-chamber PPM provides an advantage and is used in children <10 kg weight.[7] Epicardial (surgical) leads are favoured in infants and small children because of somatic growth. With the recent advancement of steroid eluding leads, epicardial leads have a better threshold and less complications.[8] Since our patient weighed 2.6 kg, we used a single chamber PPM with epicardial lead based on the evidence available in the literature. To our knowledge, our case is the smallest infant to have PPM implantation in Pakistan as recorded in literature. Previously published case series had the lowest weight of 4 kg. Post-operative complete heart blocks are managed with dual chamber PPM implantation because it is easy to place in previous sternotomy and has better accessibility to right atrium and ventricle.[9] This was not the situation in our case.

The surgical approaches available for epicardial lead placement are median sternotomy, lateral thoracotomy, left subcostal, and xiphoid approach. While traditionally, median sternotomy is favoured because of good visual and easy approach, xyphoid approach has been shown to produce equally good results with better cosmetic appearance and lesser morbidity of pain and hospital stay duration.[6] We also used the xiphoid approach in our patient.

Some of the concerns with PPM are lead dysfunction, finite battery life, and device related infections.[3] The average battery life is different between adult and paediatric populations because of the relatively higher heart rate in the latter. A study has reported the mean longevity of the pacemaker generator as 5.5 years and lead as 10.8 years in children.[10] A secondary operation is required when the battery life is completed.

Conclusion

Congenital heart block is life threatening. PPM is mandatory treatment of CHB. In low-birth-weight neonates and infants, single chamber PPM with epicardia lead offered a chance of survival. Therefore, we recommend managing small infants with this technique.

Disclaimer: This manuscript has not been previously presented or published in any conference. Author Naela Ashraf is currently affiliated with Aga Khan University Hospital and was previously affiliated with Liaquat National Hospital at the time of case report.

Conflict of Interest: The ethical review statement is signed by the co-author of this manuscript.

Source of Funding: None.

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