CASE REPORT

# Anesthetic management of a 2-day-old with complete congenital heart block

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## ABSTRACT

Maternal connective tissue disorders such as Systemic Lupus Erythematosus (most common), Sjogren's syndrome, mixed connective tissue disorders may lead to the rare condition of complete congenital heart block in the neonate. Rare fetal syndromes such as myocarditis, 18p syndrome, mucopolysaccharidoses and mitochondrial diseases are other causes. The mortality rate of this condition is inversely propotional to the age of presentation being 6 % in the neonatal age group. As the cardiac output in the neonate is heart rate dependent, it is crucial to maintain the heart rate in these patients. Pharamacological interventions with dopamine, isoprenaline, epinephrine and atropine are known for their variable response. Although permanent pacing is the most reliable mode of management, the access to it is often not readily available, especially in the developing countries. In such cases temporary pacing methods become lifesaving. Of all the modalities of temporary pacing (transcutaneous, transesophageal and transvenous) transcutaneous pacing is the most readily available and immediate mode. In this case report we present a two day old neonate with isolated complete congenital heart block and a resting heart rate of 50-55/min in immediate need of palliative surgery for trachea-esophageal fistula (TEF). With pharmacological intervention the heart rate could only be raised to 75-80/min. The surgery was successfully carried out using transcutaneous pacing to maintain a heart rate of 100/min.

**Key words:** *Anesthetic implications, complete congenital heart block, transcutaneous pacing*

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## INTRODUCTION

Congenital complete heart block (CHB) is a rare condition with incidence of 1 in 22,000 live births. The cases associated with structural heart disease have higher morbidity and mortality.[1] The mortality of congenital heart block increases as the age of diagnoses decreases with that being 43%, 6% and 0% in fetal, neonatal and childhood age group.[2] In this case report, we discuss the anesthetic management of a 2-day-old with complete congenital heart block, trachea-esophageal fistula (TEF), anorectal malformation and perforation, who presented for TEF repair and treatment of intestinal perforation.

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## CASE REPORT

A 2-day-old-male baby, weighing 3 kg, born at full term by normal vaginal delivery presented to our facility with complaints of absent anorectal opening, abdominal distention, drooling of saliva and tachypnoea. On examination, he was found to have bradycardia with a heart rate (HR) of 65-75 beats/min. The electrocardiogram (ECG) performed for evaluation of bradycardia revealed CHB [Figure 1]. On the evaluation of the mother, she was diagnosed to have syphilis, which was considered as the probable cause of the congenital heart block.

Clinical examination revealed a lethargic neonate with abdominal distention and drooling from the mouth. The child was maintaining saturation of 85% on room air. On auscultation, the new born was found to have a HR of 50-55 beats/min with bilateral crepitations in the chest. The respiratory rate was 65/min. He was intubated with 3.5 mm internal diameter endotracheal tube using the midazolam 0.3 mg and mechanically ventilated. There was no hypothermia. The lab investigations revealed a hematocrit

of 50%, platelet count of 3.2 lakh, total leukocyte count of 13,500, potassium 5.2 meq/L, creatinine of 1 mg/dL, bicarb of 20.5 meq/L, urea 30, Ca 4.5 meq/L, glucose of 80 mg/dL. An ECG was done, which showed wide QRS complexes and complete dissociation between the p-waves and QRS complexes. The echocardiography revealed a structurally normal heart with a small patent ductus arteriosus. The pediatric cardiologist diagnosed the new born to have a congenital CHB, advised to start dopamine (at 5 μg/kg/min) and isoprenaline (at 2 μg/kg/min) infusions were started to maintain the cardiac output. This increased the HR to about 75-80/min. No signs of cardiac failure were present in this neonate at this time.

The child was shifted to the operating room and attached to the ventilator. Oxygen and air were started. All the routine monitors were applied [Figure 2]. Isoprenaline and dopamine were continued. Adult size external adhesive pacemaker pads were applied antero-posteriorly and pacing was started in VOO mode with the pacing rate set to100 beats/min. The current was increased until a capture beat was detected. The threshold current was 60 mAmp. The patient was induced with fentanyl 4 μg, sevoflurane and atracurium 1.5 mg. The endotracheal tube was replaced with a rigid bronchoscope [Figure 3]. The TEF was identified and fogarty catheter was used to exclude the fistula. This was followed by palliative gastrostomy and sigmoid colostomy to relieve the intestinal obstruction. The vitals remained stable throughout the surgery with $SpO_2$ fluctuating between 85% and 92%.

## DISCUSSION

Complete congenital heart block in a neonate is a rare, but the life-threatening condition. The most common cause of congenital heart block responsible for 70-90% of the cases is neonatal lupus secondary to maternal systemic lupus erythematosus.[3] Other maternal connective tissue disorders such as Sjogren's syndrome and mixed connective tissue disorder also may lead to this condition in the neonate [Table 1]. Fetal myocarditis, 18p syndrome, mucopolysaccharidosis and mitochondrial diseases are among the rarer causes.[4] Antibody positivity in these cases is associated with a worse prognosis.[5] Congenital heart block may be associated with structural heart diseases such as left isomerism and congenitally corrected transposition of great arteries, in which case, the survival rate decreases, the 1 year survival rate being 19% in contrast to isolated congenital heart disease where it is as high as 75%.[6] Acquired atrioventricular (AV) block in children is due to traumatic obstruction to AV conduction pathways secondary to cardiac surgery.[7] The earlier the presentation of the heart block the higher the mortality

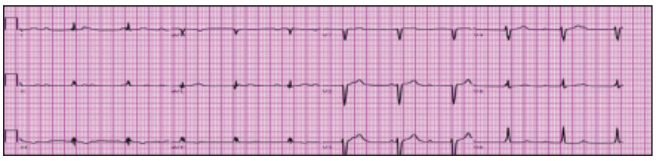

**Figure 1:** Electrocardiogram

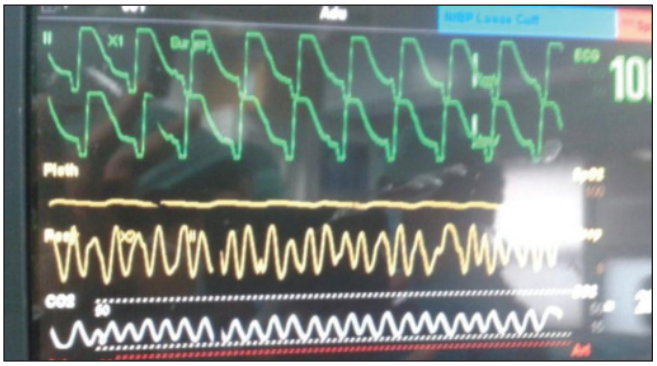

**Figure 2:** Electrocardiogram, plethesmography and end tidal $CO_2$ recordings of the patient

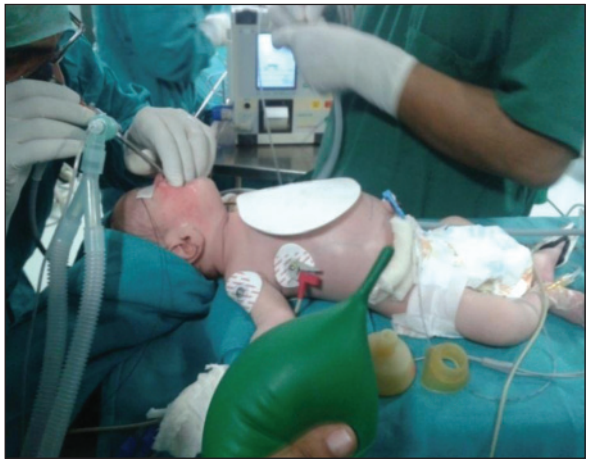

**Figure 3:** Bronchoscopy for exclusion of fistula

**Table 1: Causes of heart block in neonate**

| Congenital | Acquired |
|---|---|
| Connective tissue disease in mother | Traumatic CHB post-cardiac surgery |
| Systemic lupus erythematosus | |
| Mixed connective tissue disorder | |
| Sjogren's syndrome | |
| Fetal myocarditis | |
| 8p syndrome | |
| Mitochondrial diseases | |
| Mucopolysachhridoses | |
| Congenital heart disease | |
| Heterotaxy with AV canal defects | |
| L-transposition of great arteries | |
| Idiopathic | |

AV: Atrioventricular; CHB: Complete heart block

with fetal mortality varying from 43 while diagnosis and adequate management in the neonatal period leads to 94% survival rate.[2]

The presentation of congenital heart block varies from fetal loss and hydrops fetalis in the fetal period to congestive heart failure with irritability or lethargy, hepatomegally, mottling, pallor and cyanosis in the neonatal period to congestive heart failure and symptomatic bradycardia in children. Older children can even present with asymptomatic bradycardia.[8]

The above mentioned patient had a resting HR of 50-55/min. Even after adding pharmacological agents the rate could only be increased to around 75-80/min while the narrow complexes persisted in the ECG. Hence, mechanical intervention was necessary in this case to maintain the cardiac output. The cardiac output of a neonate is HR dependent; hence, the dire need to increase the HR in the current case. The pharmacological methods such as isoprotenerol, dopamine, epinephrine and atropine [Table 2] produce a variable and unpredictable HR response.[9] The drugs that are known to either depress AV conduction or have sympatholytic or parasympathomimetic effects should be avoided in such cases. The class 1 indications for pacing as listed by American Society of Anesthesiologists-North American Society of Pacing and Electrophysiology include; a ventricular HR <55 or <70/min with congenital heart disease, presence of wide QRS escape rhythm, complex ventricular ectopy or ventricular dysfunction, age >1 year with an average HR <50/min, abrupt pauses in ventricular rate that are 2 or 3 times the basic cycle length or associated symptoms due to chronotropic incompetence.[10]

A number of pacing modalities are available [Table 3]. Permanent pacing may often not be readily available, especially in resource limited settings. In such settings temporary pacing may become lifesaving, especially when emergency surgery for other, treatable life-threatening conditions is required, which cannot be put on hold awaiting the implantation of a permanent pacemaker. Immediate access to a means of increasing the HR either with transcutaenous pacing, or pharmaceutical therapy such as atropine and isoprenaline should be available. The available forms of temporary pacing include transcuatneous, transesophageal and transvenous. Transcutaneous pacing provides immediate pacing and is indicated and lifesaving in neonates with congenital heart blocks. Although in a recent case series transvenous pacing has been used successfully to pace neonates,[11] it is generally contraindicated in this patient group due to small body size, probable right to left heart shunt as well as possible of venous obstruction due to small size of veins.[12]

The transcutaneous pacemaker should be placed and its capture threshold and pacing efficacy should be checked prior to the initiation of surgery. Once the pacing wires are placed and the cables connected, the pacemaker should be attached and pacing commenced. VVI is the preferred mode for pacing except in cases where elctrocautery is used. In such cases VOO mode is preferred. The initial rate is determined by the resting ventricular rate of the new born and is usually kept in the low hundreds. The rate is optimized based on clinical indices of cardiac output such as the pulse and blood pressure. The current is gradually increased till the electrical and mechanical capture is detected. The size of paddles does not determine the energy needed to pace the heart. Although the current needed to pace the heart increases as the paddle size increases, the current density and hence the discomfort experienced by the patient decreases.[13] Although we had chosen the adult sized electrodes to pace the heart, it did not alter the outcome of our patient. A back up pacemaker, with additional batteries should be readily available.

**Table 2: Pharmacological methods of pacing**

| Drug | Dosage |
|---|---|
| Atropine | 0.02 mg/kg min: 0.1; max: 0.5 |
| Epinephrine | 0.01 mg/kg |
| Dopamine | 2-20 μg/kg/min |
| Dobutamine | 2-20 μg/g/min |
| Isoproterenol | 2.050 μg/kg/min |

**Table 3: Pacing in a neonate**

| Temporary pacing | Permanent pacing |
|---|---|
| Invasive | Epicardial |
| Transvenous pacing | Endocardial |
| Non-invasive | |
| Transcutaneous pacing | |
| Transesophageal pacing | |

## CONCLUSION

Complete congenital heart block is a rare, but life-threatening condition. Such neonates often have other surgical issues for which urgent surgery at birth may be required. In such cases transcutaneous pacemaker can be a good choice, since waiting for the placement of permanent pacemaker might worsen the child's condition. It is important to preoperatively test the pacemaker and make sure that it is in working condition, the preferable mode for the neonate is VVI, with the initial rate of low hundreds. A back up pacemaker and chronotropic agents such as isoprenaline and dopamine should be kept ready.

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