CASE REPORT

Transvenous Permanent Pacemaker Implantation in 14 Month Old baby with Congenital Complete Heart Block

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Abstract

Congenital Complete Heart Block in utero has been diagnosed more frequently with the clinical use of fetal echocardiography. The fetus with complete heart block may remain asymptomatic or develop congestive heart failure. Pacemaker therapy in children involves unique issues regarding patient size, growth, development and possible presence of congenital heart disease. Historically, epicardial pacing was more common in children. As technology has improved, generators and leads have become smaller and more advanced, allowing transvenous pacing system in children. Pacemaker therapy is even technically feasible in infants and neonates. We present a 14mth old baby of just 7.5kg who presented to us with Adams Stokes and subtle signs of congestive heart failure that after stabilization was implanted a transvenous MRI compatible permanent pacemaker (VVIR).

Key Words

Congenital Complete Heart Block (CCHB), Electrocardiogram (ECG)

Introduction

The incidence of Congenital Complete Heart Block (CCHB) is one case per 25,000 live births and one case per 250 births with congenital heart disease out of which L- TGA is the commonest associated anomaly (1). The ventricular rate at rest may vary from 40 to 100 but is usually between 50 to 70. The patients with congenital complete heart block are usually asymptomatic and able to achieve a normal life expectancy unless they have a severe underlying cardiac anomaly, neonatal rate of less than 50, family history of AV delay, broad n bizarre QRS complex or Adams Stokes. The incidence of associated cardiac anomalies is only 25% and mortality rate in this group is 50% more due to the severity of associated lesion than to heart block itself (2).

Pediatric pacemaker implantation is performed primarily to treat abnormalities of SA/ AV node dysfunction, congenital long QT syndrome, cardiomyopathy etc. For some time, epimyocardial pacing was the most common pediatric pacing application. At present epicardial pacing is used primarily when transvenous pacing is contraindicated or for patients undergoing concomitant heart surgery(3). Contraindication to transvenous pacing include prosthetic tricuspid valve, intracardiac shunts, recurrent transvenous lead dislodgement and probably, minimal patient size. Advantages of epicardial implantation include absence of a need to provide vascular continuity with cardiac chambers and avoidance of concomitant venous thrombosis. Disadvantages include more frequent sensing and capture failure, higher rates of insulation and conductor fracture and need for an open chest approach. However, the easier placement of transvenous leads in small patients does not imply that this approach is necessarily superior to others (4).

Case Report

A 14 month old baby weighing 7.5 kg presented to us with a history of recurrent loss of consciousness, and at the time of admission was deeply cyanosed with severe respiratory distress (*Fig 1*). On examination, the baby was drowsy with a heart rate of 30- 35 / mt, tachypneic with acidotic breathing and respiratory rate of 55/min. Chest showed bilateral crepts and cardiac examination revealed a displaced cardiac apex outwards and downwards alongwith a harsh pan systolic grade IV/ VI murmur with maximum intensity at apex. The baby was intubated, oxygenated and mechanically ventilated. On investigation, complete blood count, renal function test, liver function test and ultrasound abdomen was normal.

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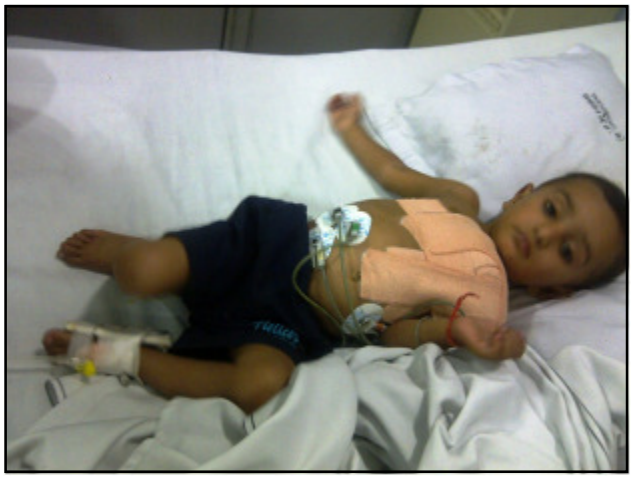

Fig. 1 14 Month Old Baby Just Weighing 7.5kg After Transvenous Permanent Pacemaker Implantation

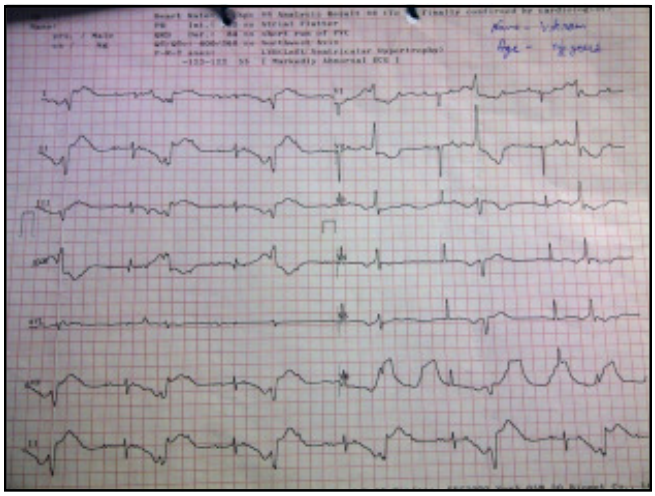

Fig. 2 An ECG Showing Complete Heart Block with Intermittent wide and Bizarre QRS Complexes

ABG showed mild metabolic acidosis with hyperkalemia. Chest X ray showed an increased cardiothoracic ratio with hyperaemic lung fields and normally located aortic arch. ECG showed a complete heart block with wide n bizarre QRS complexes in between (*Fig 2*). An echocardiography showed situs solitus, no intra or extra cardiac shunting, normal venous drainage and arterial outflow, globally dilated left ventricle with moderate mitral and severe tricuspid regurgitation with an ejection fraction of 54% (*Fig 3*). The diuretics in form of furosemide and cocktail for hyperkalemia were given intravenously alongwith antibiotics and external pacing was started. After decongestion and adequate diuresis, the baby was shifted to cath lab for transvenous permanent pacemaker implantation. UAAP, local anaesthesia and intravenous

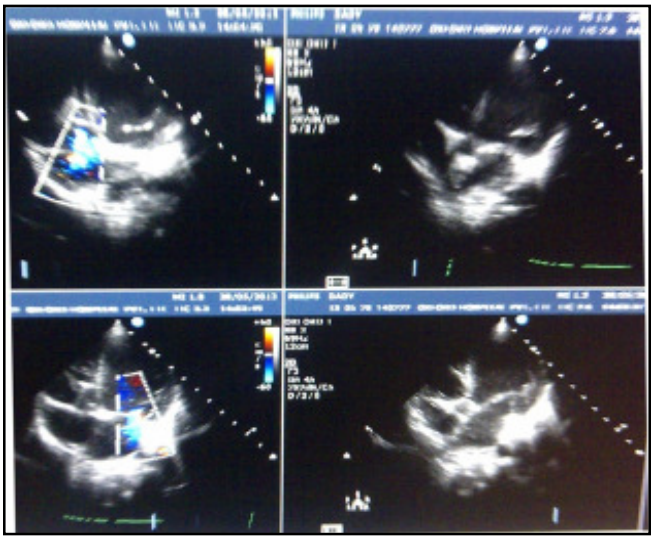

Fig. 3 An Echo Showing Severe Tricuspid Regurgitaion(3a), Moderate Mitral Regurgitation(3c), Normal Aortic Arch(3b) and no Intracardiac Shunt (3d)

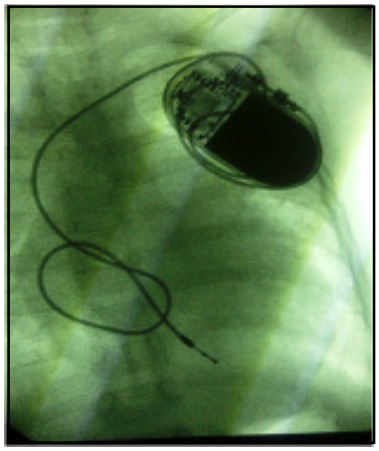

Fig. 4 VVIR MRI Compatible Permanent Pacemaker Implanted with Alpha Loop of Lead in Right Atrium

ketamine, percutaneous subclavian vein puncture to assess the venous system was made. The wire was positioned in the right heart and then after serial dilatation a 6F MRI compatible VVIR pacemaker lead was positioned in the right ventricle. A generous amount of slack in lead was kept in the right atrium to allow uncurling thereby reducing the risk of lead fracture/ dislodgement with linear growth. Testing was performed using cables to a pacing system analyzer, which ascertained an adequate sensing of intrinsic waves, capture threshold and lead impedance. The generator was then connected to the lead and pocket was fashioned submuscularly with blunt dissection and cauterization. The generator was placed in the pocket and incision was closed in multiple

layers (*Fig 4*). The pacing rate was set at 90 bts/ mt. The baby was discharged on 3rd day and is doing well on regular follow up.

Discussion

The first case report of congenital complete heart block was by Schuster in 1896. Electrocardiogram documentation was first reported in 1908 and in 1921 the first case was reported with ECG documentation at birth (5). Although a congenital etiology of heart block cannot be established definitely within intrauterine or neonatal ECG data, block which is documented electrocardiographically under the age of 2 is usually considered to be congenital provided there is no history of Congenital Syphilis, Diptheria, Mumps or Rheumatic Fever (6). Since the slow rate of congenital complete heart block during the neonatal and infancy periods has proved to be surprisingly easy to overlook, physician are urged to suspect heart block in all newborn infants and small children with heart rate less than 90.

In patients with uncomplicated congenital complete heart block, a 5 to 10% incidence of Adams Stokes episodes has been reported during infancy to childhood. The incidence of fatal ones is unclear but fatalities for the first two attacks are rare and for this reason patients usually are not considered for permanent pacemaker therapy until they have had 2 or more such episodes (7).

In a newborn with congenital complete heart block but a structurally normal heart, an epicardial ventricular pacing system initially suffices to meet hemodynamic needs. In contrast, a newborn with significant structural heart disease and congestive heart block may benefit more from a dual-chamber system to obtain AV synchrony and meet hemodynamic needs (8). In general, the transvenous route is a reasonable approach for children weighing at least 10 kg, although other authors have reported successful transvenous pacing in neonates without complications. Physical considerations include absence of intracardiac shunting, low-flow states, and anatomic barriers (e.g., mechanical valves that block pacing leads). However, because of continued growth and vigorous activity, pediatric patients have lead fracture and failure rates distinctly higher than adults (9).

Since the incidence of symptoms and death is low in CCHB, it is important to realize that the majority needs no form of therapy. Hemodynamic and cath studies have shown virtually normal resting and exercise cardiac output and physical working capacity inspite of low ventricular rate (10).

Conclusion

Fetal CHB cannot be diagnosed by ECG through mother's abdomen. However, in labour with head engaged, an electrode can be placed in the fetal scalp to make a definite diagnosis of CHB. With the recent advent of fetal echo, the atrial and ventricular rates and their relationship can be determined to diagnose CHB (11). Current practice suggests that transvenous pacing leads routinely can be placed in children who weigh more than 10 kg. As pacing technology continues to reduce lead body diameter, it is likely that children with even smaller body weights can be considered for transvenous placement. It is important to remember that pacemaker equipment presently available is far from ideal for infants and small children. A miniature system with wires and with a permanent (atomic) power source is needed.

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