Case Report

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Cardiac Syncope in a 3 Year Old: A Rare Entity

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Abstract

Complete heart block in children is a rare entity. A study published in 1998 calculated the mean incidence of isolated Complete Heart Block during 1970–1994 to be 1:17,000. This is a case report of a 3-year-old who presented with syncope, diagnosed with complete heart block and managed with a permanent pacemaker implantation.

Keywords: Congenital Heart Block; Syncope; Dual Chamber Pacemaker.

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Background

M qorquiol first described congenital complete atrioventricular block (CCAVB) in 1901 [1]. A study published in 1998 calculated the mean incidence of isolated Complete Heart Block during 1970–1994 to be 1:17,000 [2]. The overall mortality in patients without structural heart disease has been reported to be 5% to 8% [1]. The 3 pathologic processes that cause congenital heart block are lack of communication between the atrial musculature and the more peripheral part of the conduction system, interruption of the atrioventricular (AV) bundle and pathologic changes in an aberrant conduction system [1]. Congenital complete heart block can be associated with maternal metabolic diseases, drugs, viral myocarditis, transference of maternal autoantibodies,and genetic channelopathies [2].

Autoimmune congenital heart block (CHB) is an immune-mediated acquired disease that is associated with the placental transference of maternal antibodies specific for Ro and La autoantigens. The disease develops in a fetal heart without anatomical abnormalities that could otherwise explain the block and illustratescongenital heart block due to inflammation and fibrosis of the conduction system [2].

One of the first cases of congenital complete heartblock in Africa was reported by Okoroma

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et al. in Nigeria in a 3-month-old infant who was successfully treated with pacemaker implantation [4]. In Uganda, Namuyonga *et al*. reported a case of isolated CHB in an infant diagnosed at the age of 7 months and successfully treated with a permanent pacemaker. In another report, Dey *et al*. reported a case of isolated CHB diagnosed in utero at 24 weeks of gestation [4]. Paediatric pacemaker implants comprise less than 1% of all pacemaker implants [5]. We describe the management of a 3-year-old patient who presented with complete heart block.

Case Report

A 3-year-old baby weighing 8 kg presented with 2 episodes of collapse while playing at home was presented to our cardiac clinic. There was no associated convulsions or loss of sphincter control. He had normal milestone. On examination he was noted to be bradycardic with a heart rate of 39 beats per minute. 12 lead ECG done confirmed complete heart block. (Fig. 1) Echocardiogram was normal. A 24-hour-holter was also performed.

Discussions between the paediatric cardiologist and cardiothoracic surgeon concluded with a decision to put a permanent dual chamber pacemaker to avoid sudden cardiac death.

Under General anaesthesia, via right thoracotomy approach, a dual chamber permanent pacemaker with bipolar leads were placed on the right atrium and ventricle. The pacemaker generator was placed in the right hypochondriac region. The leads were left loose in order to allow for adjustment according to his growth. (Fig. 2)

Pain was managed with intercostal nerve block and local infiltration. Right sided intercostal drain was left in situ. He was extubated in theatre, observed in HDU for 24 hours and discharged home after 2 days.

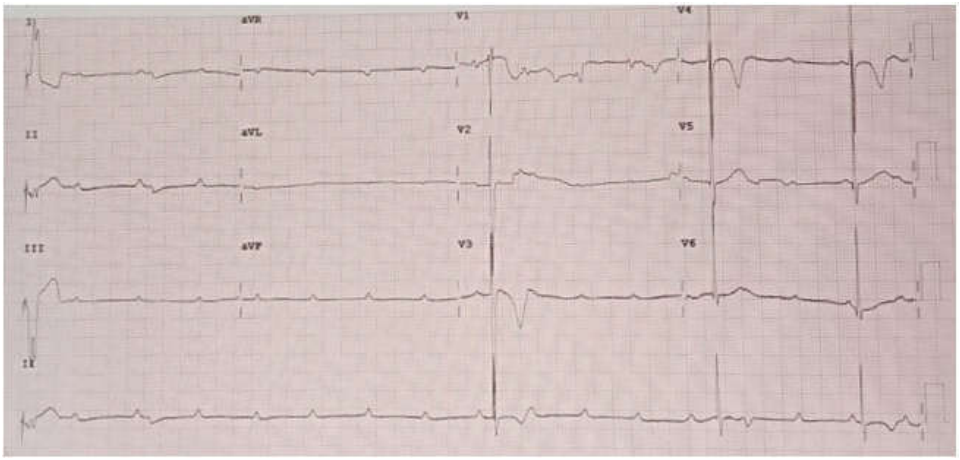

Fig. 1: Electrocardiogram of complete heart block

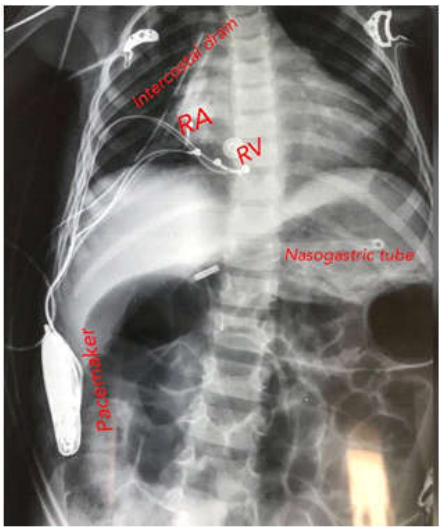

Fig. 2: Radiograph of pacemaker generator in right hypochondriac region and leads to right atrium and ventricle.

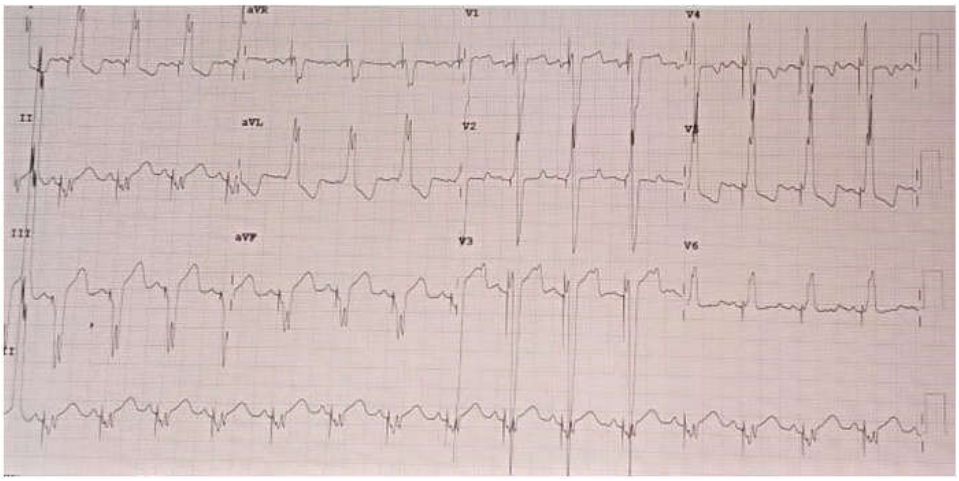

Fig. 3: Electrocardiogram of atrial sensed ventricular paced complexes

Discussion

Indications for pacing in patients with Congenital Complete Atrioventricular Block include a heart rate <55 beats/min in a neonate or <40 beats/min in a child or adolescent. Associated congenital heart disease, symptomatic bradycardia or exercise intolerance, pauses >3 seconds while awake or >5 seconds while sleeping [1]. In our case report the child had symptomatic bradycardia and exercise intolerance and a heart rate of 39 leading to the decision to implant a permanent pacemaker.

From out literature review, AV block was diagnosed at 20–24 weeks of gestation, and in 75% was during weeks 20–29. Only 2% of reported cases were diagnosed at birth or within the neonatal period [2]. Therefore, it is an uncommon occurrence to have the diagnosis made at 3 years old, as presented in this case report. A properly performed pregnancy follow-up with serial echocardiograms could aid in antenatal diagnosis and plan perinatal management when appropriate in order to optimize outcome. This emphasizes the clinical value of high quality antenatal care and proper screening [4]. But the cardiac evaluation of fetuses with suspected complete heart block may be challenging most especially in resource limited settings like ours where there is scarcity of skilled personnel that can perform fetal echocardiography [4].

A significant number of babies with complete heart block have an autoimmune aetiology. Of these, two-thirds of babies with autoimmune complete heart block required pacemakers, almost all during the first year of life, and in nearly two-thirds of cases during the first 10 days after birth [2]. Women who have babies with complete heart block of any type, should be tested (with commercial tests) for the complete panel of anti-Ro and anti-La antibodies [2]. Clinical diagnosis of autoimmune disease was present in 12 (75.0%) mothers. Among them, eight (50.0%) tested positive for systemic lupus erythematosus and four (25.0%) had Sjögren's syndrome [3].

Implantation of a dual-chamber epicardial pacemaker in neonates with congenital heart block is technically feasible and results in excellent outcomes in patients with structurally normal hearts. System longevity at 6 years is excellent [5]. Small vessel size and associated intracardiac defects make Trans venous implantation difficult or impossible. The disproportion between the small body size and the device dimensions prevents placement of the pulse generator in the chest wall. Furthermore, the effects of growth on the leads and on the lead-myocardial junction result in a high incidence of exit block and lead fractures [3].

Conclusion

This case highlights certain challenges that require tobe addressed. It is possible that this diagnosis may have been made in utero. Screening of mothers for connective tissue disease is required, especially if further pregnancies are planned. Treatment is possible leading to a return to normal life for the child.

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