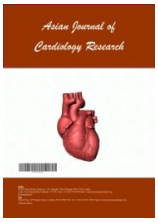

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Complete Congenital Atrioventricular Block in a Nigerian Neonate of a Previously Undiagnosed and Asymptomatic Mother with Systemic Lupus Erythematosus: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Congenital atrioventricular block is a rare disorder with an incidence of 1 in 22,000 cases. Its diagnosis is devastating to parents anywhere in the world, and even more so in a resource limited country like in Nigeria. We report the case of Baby E. who was first suspected to be having a complex congenital heart disease in utero at 26 weeks of gestation. He was delivered to an apparently healthy 25-year primigravida who was later found to have serological features of systemic lupus erythematosus (SLE). We also highlight the challenges and proffer recommendations for management of congenital atrioventricular block in Nigeria.

Keywords: Congenital; atrioventricular block; neonate; systemic lupus erythematosus; Nigeria.

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KEY MESSAGE

Congenital Complete Atrioventricular block is associated with the SLE/ Sjogren syndrome in almost 80% of cases and up to half of the mothers may have been asymptomatic. There is a rising trend of rheumatologic diseases in Nigeria even in children, and thus, there is need for capacity building in the management of congenital complete atrioventricular block in a resource poor setting like ours, close monitoring, early diagnosis, provision of sources of funding and availability of appropriate-sized pacemakers may lead to optimal outcomes.

1. INTRODUCTION

Congenital atrioventricular block (CAVB) is an arrythmia, present in utero or at birth. It is due to an abnormality in the fetal conduction system in which there is slow or lack of transmission of impulses from the sinoatrial node to the ventricles [1]. Third degree AVB is the commonest type [2]. CAVB occurs in 1 in 22,000 livebirths, however the incidence is thought to be higher because of the high rates of fetal loss associated with the condition. CAVB can be isolated or associated with structural heart disease [3].

The commonest cause of CAVB, which accounts for 80% of cases are autoimmune conditions usually the systemic lupus erythematosus or the Sjogren Syndrome. Maternal IgG auto anti bodies, Anti Ro (Anti Sjogren Syndrome A Antigen-SSA) and Anti La (Anti Sjogren Syndrome B-SSB), are known to cross the placenta and bind to the cardiac conduction tissues of the fetus [4]. The antigen antibody complex is said to evoke an immune reaction which leads to inflammation and subsequent fibrosis with damage to the sino-atrial, atrioventricular nodes or other fibres of the cardiac conduction system [4,5].

The auto antibodies are also known to affect the fetal skin, blood and liver cells. Since these cells can regenerate, their affectation is reversed once the antibodies clear by 6 – 8 months of postnatal life. Third-degree AV block is usually irreversible [6]. These cardiac and non-cardiac manifestations caused by the auto antibodies comprise the neonatal lupus syndrome.

Another cause of CAVB is the in association with complex congenital heart diseases in which there is an abnormality in the development of both the heart structurally as well as the conduction tissues.

Presentation is usually in utero most frequently with fetal bradycardia at which time the block is at least second degree. It may also present as hydrops fetalis. The newborn may be asymptomatic despite a heart rate of less than 100 beats per minute or may present with excessive sleepiness, poor feeding, features of congestive cardiac failure. Treatment is via cardiac pacing done when the patient is hemodynamically unstable, when heart rate is consistently below 55 beats per minute to mention a few [4].

2. CASE REPORT

The patient is a newborn delivered to a 25-year-old primigravida via elective Caesarean section at 37 weeks of gestation on account of fetal bradycardia and a fetal anomaly scan suggestive of a three chambered heart done at 26 weeks gestation. At 26 weeks of gestation, following a routine obstetric scan, fetal heart rate was noticed to be less than 60 beats per minute. Mother thereafter had a fetal anomaly scan in which the radiologist confirmed the fetal bradycardia and a suspected diagnosis of a three chambered heart.

Mother was referred for paediatric cardiology review at 33 weeks of gestation. There was no history suggestive of autoimmune disease; no history of joint pains, light sensitive skin rashes, recurrent fever, recurrent pregnancy loss or hair loss. She was not diabetic or hypertensive. No family history of congenital heart disease, or sudden cardiac death. On examination fetal heart rate was 60 beats per minute. Investigations including Anti Ro (SSA), Anti La (SSB), Anti nuclear antigen, Anti double-stranded DNA and erythrocyte sedimentation rate as well as fetal echocardiography were requested. Mother however defaulted following first review and was next seen after delivery of the baby.

Baby cried immediately after birth with no cutaneous lesions. Apgar scores were 8 and 10 at the first and fifth minutes respectively. He was pink in room air. Oxygen saturation was between 89 - 95% in room air, respiratory rate of 50 cycles per minute, heart rate of 44 beats per minute. The first electrocardiogram done revealed atrial

flutter with complete heart block as shown in (Fig. 1). Subsequent ECGs done did not show the flutter waves as seen in (Fig. 2). Echocardiography done revealed a moderate secundum ASD, 9 mm in diameter and a tiny PDA.

The newborn developed tachypnea, displaced apex to the 5th left intercostal space and bilateral pitting pedal oedema at one week of life. He was managed for congestive cardiac failure with intranasal oxygen. Investigations done in the newborn include anti Ro, Anti la, normal liver function tests and a full blood count which was normal except for mild thrombocytopaenia of 142,000 cells/mm [3]. Mother's anti Ro antibody screen was positive with a titre of >240 U/ml while anti La was negative. Anti la with a titre of the baby was >240 U/ml, Anti Ro was negative. Baby was planned for an epicardial cardiac pacemaker. Neonatal sized pacemakers were not available in our center, despite several attempts to reach the suppliers. Parents were however also reluctant to have a "battery-powered child". Parents could also not afford to pacemaker insertion due to financial constraint. Baby was on follow up two-weekly in clinic, but they later defaulted. Baby was brought in dead to the emergency room at three months of age after a febrile illness that lasted 48 hours.

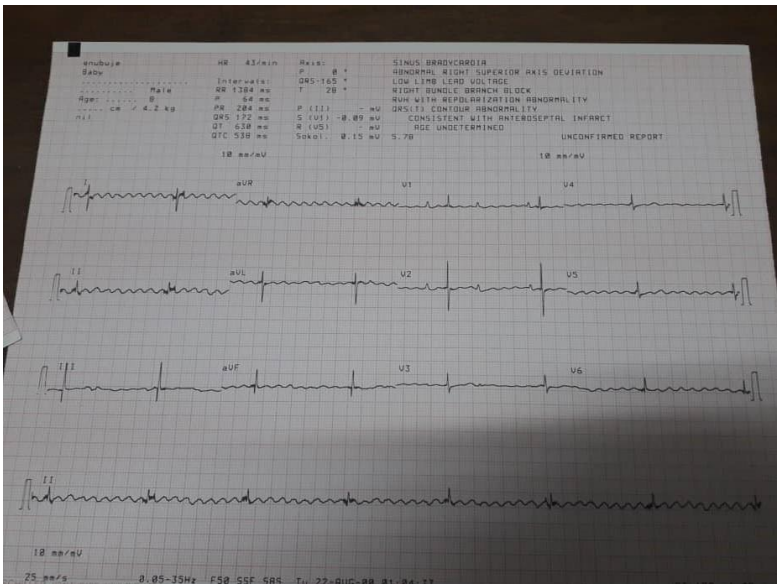

Fig. 1. Initial ECG tracing showing complete atrioventricular block with atrial flutter

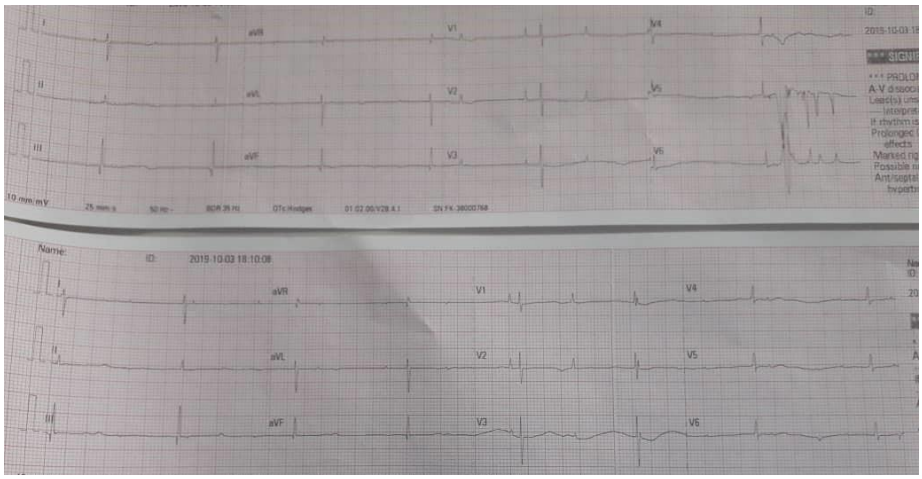

Fig. 2. Second ECG showing complete atrioventricular block without flutter waves

3. DISCUSSION

There are only a few case reports of CAVB in Nigeria [7,8]. CAVB is most commonly associated with SLE/autoimmune conditions even though 50% of the mothers are usually asymptomatic and undiagnosed as was the case with our patient. The risk of occurrence of CAVB in a mother with anti Ro/La positive is about 3% while the risk of recurrence in subsequent pregnancies is almost 20%. Experimental studies have shown that fetal tissues between 18 – 24 weeks rather than adult tissues have an abundant of anti SSA(Ro) antigens [9]. This explains why anti Ro, rather than anti La is more implicated in CAVB. This also helps us understand why maternal cardiac tissue is spared [10].

Bradycardia was first noticed in our patient at 26 weeks, which is right after the period of likely damage by the antibodies. Some foetuses present with hydrops signifying severity and may lead to preterm delivery which carries a worse prognosis. As in our patient most mothers are first diagnosed of SLE following diagnosis of an offspring with congenital heart block [5].

Interventions to prevent development of CAVB in autoantibody positive mothers include close monitoring with fetal echocardiograms as well as use of steroids, plasmapheresis and of intravenous immunoglobulin (IVIG). There is no evidence that these agents prevent onset of CAVB, but dexamethasone has been shown to halt progression of the AV block to third degree which is irreversible. It is recommended that SLE/Sjogren—syndrome patients have weekly fetal echocardiograms to measure mechanical PR intervals from 18 weeks of gestation to ensure early diagnosis of CAVB [6].

Dexamethasone has been used with variable success to slow down progression of antibody mediated fetal conductive tissue damage. Dexamethasone is thought to reduce the progression of conductive tissue damage through its anti-inflammatory properties and it's also thought to lower levels of maternal auto-antibody. It however could not be used for our patient in utero because the mother defaulted after initial diagnosis. It is however not commenced routinely because there is no evidence that it prevents development of CAVB in antibody positive pregnant mothers. Prolonged steroid use has side effects of steroids include infection, osteoporosis, osteonecrosis, diabetes, hypertension, premature rupture of membranes, preterm labour and preeclampsia for the mother and infection, adrenal insufficiency, intrauterine growth restriction (IUGR), and oligohydramnios in the fetus. In a PITCH study, the use of intravenous immunoglobulin (IVIG) in prevention of CAVB in auto-antibody positive mothers proved ineffective [11].

Treatment options in the affected newborn include watchful waiting for signs of exercise intolerance and insertion of a cardiac pacemaker. Indications for a pacemaker include very slow heart rates (below 55 beats per minute), symptoms such as poor exercise tolerance, cardiomegaly, long QRS or QT durations, ectopy, syncope, or structural or functional heart disease [3]. Unfortunately, there have been case reports of development of cardiomyopathy within the first two years of life despite early insertion of a pacemaker [12].

In our patient, signs of congestive cardiac failure developed by the first week and heart rate was consistently below 55 beats/ minute signalling the need for a pacemaker. Challenges faced were lack of a neonatal pacemaker, another issue was little experience of the cardiac team with neonatal pacing even though pacing for adults and older children are done routinely. Financial constraint was a major hurdle, because parents pay out-of-pocket and could not readily afford the investigations to be carried out and are currently sourcing for funds for pacemaker insertion. Parents also felt it was socially unacceptable for their child to be 'battery powered' and required several sessions of counselling to be favourably disposed to a pacemaker use.

4. CONCLUSION

In conclusion, the diagnosis of CAVB is heart-breaking news for the parents, CAVB is associated with the SLE/ Sjogren syndrome in almost 80% of cases and up to half of the mothers may have been asymptomatic. Prevention in auto-antibody positive mothers include serial echocardiograms from 18 weeks of gestation and use of dexamethasone in utero if any sign of CAVB is detected. A paper by Adelowo, [13] et al., revealed a rising trend of rheumatologic diseases in Nigeria even in children, and thus, there is need for capacity building in the management of CAVB. In a resource poor setting like ours, close monitoring, early diagnosis, provision of sources of funding

and availability of appropriate-sized pacemakers may lead to optimal outcomes.

CONSENT

As per international standard, parental written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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