

**Odetunde OI**  
**Ekwochi U**  
**Chinawa JM**  
**Ejim EC**  
**Ezugwu FO**

## Conducting defect with atrial septal aneurysm in a preterm neonate: A case report from resource limited setting

DOI:<http://dx.doi.org/10.4314/njp.v42i1.14>

Accepted: 7th August 2014

Odetunde OI (✉)  
 Chinawa JM  
 Department of Paediatrics,

Ejim EC  
 Department of Medicine,  
 University of Nigeria Teaching  
 Hospital, Ituku Ozalla, Enugu,  
 Enugu State,  
 Nigeria.  
 Email: odetoyintola@yahoo.co.uk

Ekwochi U  
 Department of Paediatrics,

Ezugwu FO  
 Department of Obstetrics and  
 Gynaecology,  
 Enugu State University Teaching  
 Hospital, Enugu,  
 Nigeria.

**Abstract:** Conducting defect (CD) co-existing with interatrial septal anomaly (ISA) like Atrial Septal Aneurysm (ASA) is a rare congenital heart disease presenting with bradyarrhythmia in-utero due to defects in the heart electrical conducting system. We report a case of a preterm female 'CC' (GA=35weeks) low birth weight (2.4kg) neonate with CD and associated ASA. CC's mother presented with poor foetal movements and bradyarrhythmia and had an emergency caesarian section at 35weeks with Apgar score of 3, 5. She was managed for perinatal Asphyxia. This report highlights the fact that ASA resolved

at around the first year of life but the associated heart electrical conducting system defects persisted. The report also emphasizes the challenges of managing a rare neonatal cardiac condition in a resource limited setting and the importance of thorough foetal biophysical profile and early interventional delivery when it is necessary.

**Key words:** Conducting defect; neonatal; Atrial Septal Aneurysm.

### Introduction

Conducting defect (CD) is a cardiac conduction disorder in which there is dissociation in the conduction of impulse from the sino-atrial (SA) node to the atrio-ventricular (AV) node<sup>1</sup>. This may also result in dissociation between the atrial activity (P-wave) and the ventricular activity (QRS complex)<sup>1</sup>.

It is a disorder occurring in about 1 in 22000 live births<sup>2</sup>. It has a high morbidity and mortality rate especially when associated with structural defects, common among which are; interatrial septal anomaly (ISA) like Atrial Septal Aneurysm (ASA), levo-transposition of great vessels, left atrial isomerism<sup>3</sup>. The prevalence of ASA has been reported high in the preterm neonates with high resolution rate at the end of first year of life<sup>4,4</sup>. Although ASA is considered to a benign and transient observation, reports have shown that it could be a cause of foetal atrial arrhythmia and impulse conduction disorders in adults<sup>6,7</sup>.

In the presence of a co-existing structural cardiac defects or very low heart rate in the tune of <55bpm, CD can be complicated by low cardiac output, congestive cardiac failure and death. Treatment of choice in such condition

will entail surgical insertion of a permanent pacemaker<sup>3,8</sup>. Access to this treatment option in our economy is strongly limited by cost and dearth of such service providers.

However, in the absence of the above conditions, patient with CD could generally remain asymptomatic and may need no active treatment.

This is a case report of CD in a female preterm, low birth weight neonate with associated ASA that spontaneously resolved at End of first year of life. Her heart rate has persistently remained within the range of 60-70bpm. She has remained generally asymptomatic and has gained weight progressively without insertion of a pacemaker.

We present this case because of its rarity and to highlight CD as a cause of foetal poor biophysical profile and peri natal asphyxia. A need for urgent intervention perinatally, further investigations and follow-up of such baby may be necessary.

### Case report

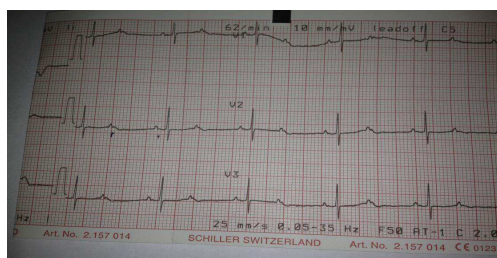
We present a preterm female baby 'CC' whose mother complained of poor fetal movements at a gestational age of 35weeks and obstetric ultrasonographic scan at same

period revealed a poor biophysical profile with a foetal heart rate of 90bpm. Following the above findings, baby was delivered via an emergency caesarean section. The APGAR score at birth was three in the first minute and five in the fifth minute. The liquor was clear. Relevant examination findings at birth include; heart rate (HR) of 112bpm regular, core temperature of 35.6<sup>o</sup>c with acrocyanosis, grunting respiration (Respiratory Rate (RR) =62cpm). The anterior fontanelle was patent and normotensive. There was poor motor activity and weak cry on stimulation. Moro reflex was incomplete and other primitive reflexes depressed. Other systemic findings were unremarkable. The birth weight was 2.4kg. The diagnosis at birth was low birth weight, prematurity with moderate perinatal asphyxia.

Initial resuscitation was done by drying and provision of warmth, suctioning to clear the air way and intra nasal oxygen therapy commenced at 1L/min. she was then transferred to the incubator set at a real temperature of 36<sup>o</sup>c. Other treatments offered include; intravenous fluid 10% dextrose in water, intravenous dexamethasone, intramuscular vitamin K, intravenous ceftazidime (antibiotic prophylaxis)

By the 3<sup>rd</sup> day of life, the HR decreased to 70bpm, oxygen saturation stabilizes at 94% without oxygen, baby has passed meconium, grunting respiration resolved, and suckling reflex was stronger. A diagnosis of bradyarrhythmia was made and she was to be investigated to determine the cause. Myocardial dysfunction secondary to perinatal asphyxia was considered as the cause and to rule out presence of congenital heart block. Electrocardiogram (ECG) was done (with improvised available adult leads) (Figure 1) and a diagnosis of conducting defect (2:1 degree block) with dissociation between the atrial activity (P-wave) and the ventricular activity (QRS complex) (Figure 2).

**Fig 1:** The ECG revealed complete dissociation between the atrial activity (P-wave) and the ventricular activity (QRS complex)



**Fig 2:** Patient with improvised adult ECG Leads



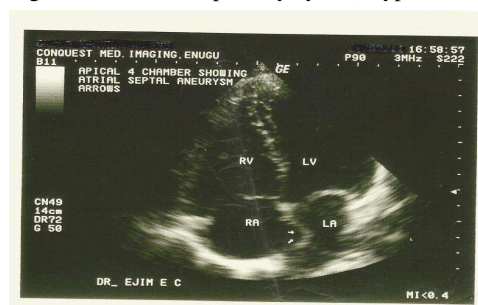
Baby continued to improve on admission and by the 9<sup>th</sup> day of life; tolerates 15mls of expressed breast milk per

feed(3 hourly). The heart rate was 64bpm regular and she remained stable. Echocardiography was done to rule out presence of any congenital structural heart defect. Mother was also screened for the presence antinuclear antibody (ANA) against systemic lupus erythematosus (SLE). Mother's serological screening tests were negative for ANA,

#### ANCA and anti ds Nase

On the 12<sup>th</sup> day of life, baby was weaned off incubator, HR was 68/min, tolerate 35mls of EBM per feed. Direct suckling was commenced. 2-D-echo cardiogram revealed ASA protruding from the midline of the atrial septum to the left atrium throughout the cardiorespiratory cycle (a type 2L ASA) (Figure 3). Two days later she was discharged home and to be followed-up on regular appointment days visit in the clinic.

**Fig 3:** Patient's Echocardiography done on the 10<sup>th</sup> day of life (Trans-thoracic Two-D-echocardiogram) revealed ASA protrudes from the midline of the atrial septum to the left atrium throughout the cardiorespiratory cycle (a type 2L ASA)



Her appointments were kept regularly and by last follow-up visit at 1 year old of age, patient has done well with a weight of 10kg. Heart rate was 60bpm. At the end of first year of life, a follow-up Trans -thoracic echocardiography (TTE) was done and revealed disappearance of the ASA (Figure 4). Baby is still on follow-up and keeping appointments and the need for pacemaker device has been discussed and prompted.

**Fig 4:** Patient's Echocardiography at the end of first year of life showing resolution of the Atrial Septal Aneurysm.



#### Discussion

There are many evidences associating Atrial Septal Aneurysm (ASA) and cardiac impulse conduction disorder<sup>6,7</sup>. Several reports have shown possible association of Right Bundle Branch Block (RBBB) and ASA<sup>6,7</sup> but few

or no data in medical literature have reported association between ASA and Congenital Conducting defect (CD).

CD is estimated to occur in one of every 22000 live births<sup>2</sup> with 25% to 33% of cases associated with other congenital heart diseases<sup>9</sup>. This rarity is also confirmed in this case, since this is the first time it is been recorded in our setting. The prevalence of ASA is said to be high among preterm neonate with high resolution rate at the end of first year of life without any complication<sup>4</sup>. The index patient is premature baby and the Trans –thoracic echocardiography (TTE) done shortly after birth revealed type 2L ASA which resolved at end of one year of patient's age with persistent of the CD. Historically, CD was thought to be a relatively benign condition, with the only significant morbidity being the need for pacemaker implantation to guard against the risk of Stokes-Adams attacks and sudden cardiac death<sup>10</sup>. This is also in tandem with our case where the patient was relatively stable with a low heart rate.

Identified risk factors in CD include the presence of hydrops<sup>11</sup>, low ventricular rate (usually defined as <55 beats/min), and prematurity<sup>12</sup>. Our patient although preterm, never had a recorded heart rate of < 55beats/min.

A study of American Lupus Registry showed 113 children with CD in which 85 of them were diagnosed in utero<sup>13</sup>. The mortality was 19%, with most deaths occurring in the neonatal period or in-utero. The only risk factor for death was delivery at <34 weeks gestation age. Our patient was 35 weeks gestation age and this may explain why she survived. It is important to note that some maternal autoimmune disease such as SLE (systemic lupus erythromatosis) may predispose foetus to CD due to auto antibodies (anti-Ro antibodies). Women with serum titers of anti-Ro antibody carry a 3% risk of having a child with neonatal lupus syndrome. If she has a prior experience with affected foetuses, her risk rises to about 18%<sup>14</sup>. The mechanism of causation of neonatal lupus is not completely understood but evidence points to the foetus beginning life with a normal cardiac structure and conducting system. Providentially, the mother of this patient was screened for lupus antibodies and she was negative.

Eronen et al reported the results of 91 infants with CD (83 diagnosed in utero)<sup>15</sup>. The mortality was 16%, with most deaths occurring in infancy. The risk factors for the mortality are low birth weight, male sex, and complications from prematurity or neonatal lupus<sup>15</sup>. Our patient is a female child with borderline low birth weight. Again this might also explain the factor why our index patient survived beyond the neonatal period<sup>15</sup>.

Significant pauses on 24-hour ambulatory electrographic monitoring may also be an indication for putting in a pacemaker. Some studies have suggested that a prolonged QTC interval or a wide QRS escape rhythm with complex ventricular ectopic may warrant urgent intervention<sup>15</sup>. Our patient did not have any of these findings.

The use of permanent pacemakers as initial pacing modality or use of temporary epicardial pacing wires in the high-risk neonate with CD is a definitive surgical treatment. Due to lack of facility, our patient had no pacing, more so she survived without much intervention, she is one year old now and had no complication. Medical management involves the use of steroid and atropine<sup>16</sup>. Atropine therapy enhances conduction through the AVN by reducing vagal tone via receptor blockade. It also improves the ventricular rate if the site of block is in the AVN<sup>17</sup>. In some cases patient may be asymptomatic and needs no intervention. This is akin to our patient. We use dexamethasone in our patient as a prophylaxis for chronic lung disease owing to the gestational age at delivery. It is noted that early postnatal dexamethasone therapy has been used recently for the possible prevention of chronic lung disease (CLD) in preterm infants with a very good outcome<sup>18</sup>. More so, the use of dexamethasone has been noted to be beneficial to children with Congenital heart block. For instance, it is documented that steroid given as 0.1 mg per kilogram to newborns with congenital heart block reduces mortality and morbidity<sup>19</sup>.

---

## Conclusion

Conducting defect especially with ASA is rare, and when coexisting, the ASA could still runs the natural course of resolution at the end of first year of life and the fact remains that in the absence of other coexisting cardiac anomaly or very low heart rate, patients with CD could thrive without a pacemaker device in the neonatal and early infant age.

<p><b>Conflict of Interests:</b> None  <b>Funding:</b> None</p>
---

---

## Acknowledgement

We wish to acknowledge the medical and nursing staff Neonatal Special Care Unit of St Patrick's Hospital, Owerri Road, Enugu, Nigeria where the baby was delivered and primarily managed.

---

## References

1. Friedman DM, Duncanson LJ, Glickstein J, Buyon JP. *Images Paediatr Cardiol* 2003;16:36-48.
2. Kertesz NJ, Fenrich AL, Friedman RA. Congenital complete atrioventricular block. *Texas Heart Inst J* 1997; 24: 301-7.
3. Friedman RA. Congenital atrio-ventricular block, pace me now or pace me later? *Circulation* 2003; 29:283-5.
4. Ozcelik N, Atalay S, Tutar E, Ekici F. Prevalence of interatrial septal aneurysm in newborns and their natural course. *Pediatr Cardiol* 2006;27:243 – 6.

5. Baysal K, Belet N, Kolbakir F, Yalm T. Atrial septal aneurysm in children. *Turk J Pediatr* 2001; 43: 293 – 7.
6. Bakalli A, Kamberi L, Pllana E, Gashi A. Atrial septal aneurysm association with additional cardiovascular comorbidities in two middle age female patients with ECG signs of right bundle branch block: Two case reports. *Case J* 2008; 1: 51.
7. Bakalli A, Pllana E, Kocinaj D, Bekteshi T, Dragusha G, Gashi M, Musliu N, Gashi Z. Association of interatrial septal abnormalities with cardiac impulse conduction disorder in adult patients: experience from a tertiary center in Kosovo. *Heart Int* 2011; 6: 11- 5.
8. Eronen M. Long-term outcome of children with complete heart block diagnosed after the newborn period. *Pediatr Cardiol* 2001; 22: 133-7.
9. Pordon CM, Moodie DS. Adults with congenital complete heart block: 25-year follow-up. *Cleve Clin J Med* 1992; 59: 587-90.
10. Waltuck J, Buyon JP. Autoantibody-associated congenital heart block: outcome in mothers and children. *Ann Intern Med* 1994;120:544- 51.
11. Schmidt KG, Ulmer HE, Silverman NH, Kleinman CS, Copel JA. Perinatal outcome of fetal complete atrioventricular block: a multicenter experience. *J Am Coll Cardiol* 1991; 17: 1360-6.
12. Eronen M, Siren MK, Ekblad H, Tikanoja T, Julkunen H, Paavilainen T. Short- and long-term outcome of children with congenital complete heart block diagnosed in utero or as a newborn. *Pediatrics* 2000; 106: 86-91.
13. Buyon JP, Hiebert R, Copel J, et al. Autoimmune-associated congenital heart block: demographics, mortality, morbidity and recurrence rates obtained from a national neonatal lupus registry. *J Am Coll Cardiol* 1998; 31:1658–66.
14. Glickstein JS, Buyon JP, Friedman D. Pulsed Doppler echocardiographic assessment of the fetal PR interval. *Am J Cardiology* 2000; 86: 236- 9.
15. Eronen M, Siren MK, Ekblad H, Tikanoja T, Julkunen H, Paavilainen T. Short and long-term outcome of children with congenital complete heart block diagnosed in utero or as a newborn. *Pediatrics* 2000; 106: 86–91.
16. Ruffatti A, Milanesi O, Chiandetti L, Cerutti A, Gervasi MT, De Silvestro G, Pengo V, Punzi L. A combination therapy to treat second-degree anti-ro/la-related congenital heart block. a strategy to avoid stable third-degree heart block?. *Lupus*. 2012;21:666-71.
17. Third-Degree Atrioventricular Block Treatment & Management. Obtainable at [emedicine.medscape.com/article/162007-treatment](http://emedicine.medscape.com/article/162007-treatment). Accessed on 28<sup>th</sup> July 2014.
18. Yeh TF, Torre JA, Rastogi A, Anyebuno MA, Pildes RS. Early postnatal dexamethasone therapy in premature infants with severe respiratory distress syndrome: a double-blind, controlled study. *J Pediatr* 1990;117:273–282.
19. Rein AJ, MevorachD, Perles DZ, Gavri S, Nadjari M, Nir A, Elchallal U. Early Diagnosis and Treatment of Atrioventricular Block in the Fetus Exposed to Maternal Anti-SSA/Ro-SSB/La Antibodies: A Prospective, Observational, Fetal Kinetocardiogram–Based Study. *Circulation*. 2009;119:1867-1872