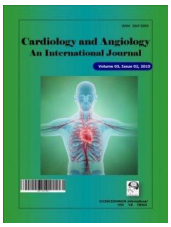

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Double Outlet Right Ventricle with Dextrocardia and Situs Inversus in a Three-Year-Old Boy: A Case Report and Review of Literature

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Authors' contributions

This work was carried out in collaboration between all authors. Author ROA developed the concept, defined the intellectual content and did part of the literature search while author AOS was involved with the data acquisition and initial manuscript preparation. Author MOO contributed to the development of the intellectual concept and editing of the manuscript. All the authors reviewed and approved the final draft of the manuscript.

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Case Report

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ABSTRACT

Aims: To highlight the rare occurrence of double outlet right ventricle (DORV) with Dextrocardia and Situs Inversus in a three-year-old boy viz-a-viz what has been reported in the literatures.
Presentation of Case: N.A is a three year old boy who presented with easy fatiguebility, bluish discolouration of lips and tongue and occasional dyspnoea on exertion noticed since about 12 months of life. There was no associated history of cough, leg swelling or frequent hospitalization. Physical examination revealed a small for age, centrally cyanosed boy with conjuctival ejection, and grade 4 digital clubbing. Pulse rate was 100 beats /min regular and synchronous with other peripheral pulses. There was a praecordial bulge to the right with apex beat at the 4th right intercostal space mid-clavicular line. Heart sounds were 1st and 2nd with a grade 2 systolic murmur at the right upper sternal border. The liver was 4 cm palpable below the left costal margin firm non-tender.
Pulse oximetry showed SPO2 value of 75-79% in ambient air. He had chest radiograph, electrocardiogram, abdominal ultrasound and two-dimensional echocardiography which confirmed a

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diagnosis of DORV with Situs totalis.
Discussion and Conclusion: Situs Inversus totalis (Dextrocardia co-existing with Situs inversus) is associated with lower incidence of congenital heart defects as compared with Dextrocardia and Situs solitus. The defects commonly associated with it, are a transposition of great arteries (TGA) and ventricular septal defects (VSD) unlike in the index patient where DORV is being reported.

Keywords: Congenital; dextrocardia; double outlet right ventricle; situs inversus.

1. INTRODUCTION

Double Outlet Right Ventricle (DORV) is a cyanotic congenital heart disease in which both the aorta and pulmonary artery arises from the right ventricle with a ventricular septal defect (VSD) being the only outlet from the left ventricle [1]. It represents <1% of all congenital heart defects.

Dextrocardia is an abnormal congenital positioning of the heart in which the heart lies in the right side of the chest as against its natural position on the left. Situs inversus is a condition in which the primary visceral organs are mirrored from their standard positions, that is the left morphological atrium is on the right while the morphological right atrium, the liver and gallbladder are on the left [2].

Situs inversus with dextrocardia is also termed situs inversus totalis because the cardiac position, as well as the atrial chambers and abdominal viscera, is a mirror image of the normal anatomy [3]. It is a rare condition with prevalence put at 1 in 10,000 in some population [5]. It is generally asymptomatic except when associated with congenital heart defects – commonly transposition of great vessels - which is seen in 5 to 10% of cases [4].

The case presented below is that of a 3 yr old boy with double outlet right ventricle, dextrocardia and situs inversus.

2. PRESENTATION OF CASE

N.A is a 3 year old Nigerian male child who presented at the paediatric outpatient clinic of our health facility with complaints of easy fatigueability noticed since 12 months of life. There was history of bluish discolouration of lips and tongue as well as occasional dyspnea on exertion but with no history of cough, body swellings or frequent hospitalizations. He is the third of 3 children and a product of term gestation delivered via caesarean section on account of antepartum haemorrhage. There was no history of exanthematous febrile illness or exposure to radiation in mother during pregnancy. Mother, however, had ingestion of herbal medications throughout pregnancy. There were no adverse events during the neonatal period. He had no feeding difficulties and was said to be otherwise well until onset of symptoms.There is no history suggestive of Consanguinity between the parents.

On examination, he was centrally cyanosed (SPO_2 of 75 - 79% in room air), had conjuctival ejection, grade 4 digital clubbing with no obvious dysmorphic features or pedal oedema. His weight was 11 kg and height was 80 cm, both are less than the 3rd percentile for age using the 2007 WHO growth chart. His peripheral pulses were regular and synchronous with a rate of 100 bpm, full volume. There was precordial bulge to the right and cardiac apical impulse was located in the fourth right intercostal space. His heart sounds were 1st and 2nd with a grade 2/6 systolic murmur at the upper right sternal border. He had a palpably enlarged liver which was 4 cm (span of 10 cm) below the left costal margin, firm and non-tender.

His haematocrit ranges between 60.9-70.1% (Hb:18.2-22.4 g/dl). Chest radiograph showed cardiac apex in the right hemithorax with oligaemic lung fields and stomach gas shadow on the right. Abdominal ultrasound scan demonstrated reversed anatomy with the liver on the left while the spleen was on the right. A 2D echocardiogram done revealed a double outlet right ventricle (DORV) with tetralogy of Fallot (TOF) physiology.

A diagnosis of DORV with Dextrocardia and Situs inversus was made and parents were counselled on the nature of the disease and available treatment options. Parents are financially incapacitated hence, for now no plan for surgical intervention but patient is on follow-up.

3. DISCUSSION

DORV has four sub-divisions based on the position of the VSD and absence or

presence of a pulmonary stenosis. They include DORV with subaortic VSD (55%-70% of cases), DORV with subpulmonary VSD (10%-30% of cases), DORV with doubly committed VSD (<5% of cases) and DORV with non-committed VSD (10% of cases) [3].

In the index patient, the echocardiographic finding was that of a large aorta overriding a large unrestrictive VSD, this describes the lesion in DORV with subaortic VSD. There was also an accompanying narrowing of the main pulmonary artery with a Peak gradient of 15 mmHg across the pulmonary valve (measures 0.73 cm), giving rise to the Tetralogy of Fallot physiology found in this patient.

DORV has been reported to co-exist with other congenital cardiac defects. Occurrence rates of associated cardiovascular anomalies include: pulmonary stenosis 21-47% (most commonly observed with subaortic type VSD), atrial septal defect 21-26%, patent ductus arteriosus 16%, atrioventricular canal 8%, subaortic stenosis 3-30%, coarctation/hypoplastic arch/interrupted aortic arch 2-45%, and mitral valve anomalies 30% [5].Our patient had the most commonly reported associated cardiovascular anomaly (pulmonary stenosis) alongside the most common form of DORV (subaortic type VSD).

There has been a previously reported case series of DORV with associated Ventricular septal defect(VSD), Pulmonary Stenosis (PS) and Atrial Septal Defect (ASD) [6] but nothing on DORV occurring with Situs inversus totalis as found in our patient to the best of our knowledge.

Clinical features of DORV depend on the variant. DORV with TOF physiology typically present with features of Tetralogy of Fallot such as hypoxic spells, cyanosis, digital clubbing and systolic murmur. All of which were present in our patient.

Dextrocardia is a congenital anomaly in which the base-to-apex axis of the heart points to the right at birth instead of the normal left direction. It was first recognized by Marco Severino in 1643 [3]. Dextrocardia is believed to occur in approximately 1 in 12,019 pregnancies [7]. A chest radiograph and ECG tracing are enough as a complement with clinical examination findings to diagnose Dextrocardia. The typical ECG findings are those of a predominantly negative P wave, QRS complex and T wave in Lead I. There could also be low voltages in V_3 –V_6 since these leads are normally placed on the left side of the chest. In the index patient, the ECG tracing showed a negative P wave & T wave in lead I as well as low voltage in leads V_3 –V_6 which are in-keeping with Dextrocardia. The Chest radiograph also demonstrated the cardiac shadow oriented to the right. These investigation results are shown in Figs. 1 and 2. The exact cause of Dextrocardia is unknown but it has been linked to several factors including autosomal recessive gene with incomplete penetrance, maternal diabetes, cocaine use and conjoined twinning [8].

Situs inversus was described about a century after Dextrocardia. It was first recognised by Matthew Baillie as a congenital defect in which there is a complete mirror-image reversal of the thoracic and abdominal organs [3]. Dextrocardia occurring with situs inversus is referred to as situs inversus totalis. It is a rare condition with an incidence of 1 in 10,000 of the general population, no racial predilection, and a male-to-female ratio of 1:1 [9]. Situs inversus is believed to occur as a result of absence of a single protein, due to mutation on murine Chromosome 12, which is important in the regulation of correct handedness [10].

Patients with dextrocardia and Situs inversus are usually asymptomatic and discovered commonly during routine examination or while being evaluated for unrelated illness as documented in several literatures [9-13]. This is in contrast with our patient who came into notice with symptoms referable to the heart.

Situs inversus totalis is known to be associated with lower incidence of congenital cardiac disease (0-10%) as compared to dextrocardia with situs solitus[8]. The cardiac defects commonly found in association with Situs inversus totalis are Transposition of great vessels and ventricular septal defects [14] as against DORV found in the index patient.

The recognition of Situs inversus totalis is important for preventing surgical mishaps that could occur from the failure to recognize reversed anatomy or an atypical history. For example, in a patient with situs inversus, cholecystitis typically causes left upper quadrant pain, and appendicitis a left lower quadrant pain.

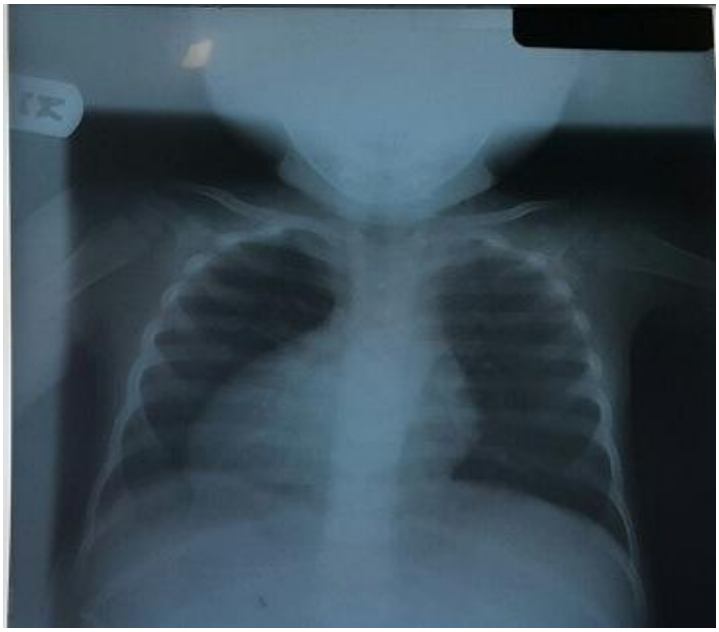

Fig. 1. Chest X-ray

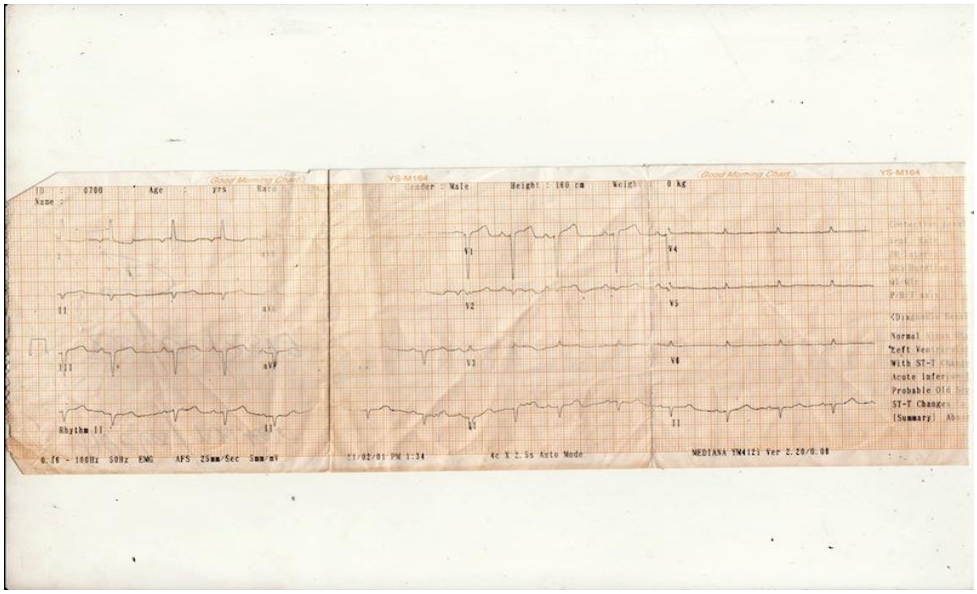

Fig. 2. ECG

4. CONCLUSION

This case highlights the rare occurrence of Double Outlet Right Ventricle with Situs Inversus Totalis which, to the best of our knowledge, has not been reported in Nigeria. This adds to the growing list of various possible cardiac anomalies that may co-exist with dextrocardia and situs inversus.

CONSENT

I wish to thank the patient, the caregiver who agreed for the report to be written and the doctors in the unit for detailed documentation.

ETHICALAPPROVAL

Approval was obtained from the health ethical committee of the Benue State University Teaching Hospital before writing the report.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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