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Case Report

DEXTROCARDIA WITH SITUS VISCERUM INVERSUS TOTALIS IN A 65 YEAR OLD MAN: A CASE REPORT

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ABSTRACT
We report a 65 year old hypertensive and type 2 diabetic low income male observed during physical examination to have right sided cardiac apex and heart sounds. Radiologic, electrocardiography, echocardiography and computed tomogram scan studies confirmed dextrocardia with situs viscerum inversus totalis. There was neither coexisting congenital cardiac anomaly nor disconcordant atrio-ventricular connection. In spite of being diabetic and hypertensive, he had no evidence of ischaemic heart disease. The blood pressure and blood glucose were adequately controlled using appropriate agents. This case, to our knowledge, may be the documented oldest Nigerian with situs inversus dextrocardia, and shows that individuals with this rare thoracic-abdominal organ developmental anomaly may have normal life expectancy similar to the general population.

Key Words: Dextrocardia, situs inversus. *(Accepted 28 October 2008)*

CASE REPORT
A 65 year old low income male Nigerian presented in the out patient medical unit of our hospital with 3 months history of orthopnoea, paroxysmal nocturnal dyspnoea, dry cough, and leg and abdominal swelling. The identified risk factors of cardiovascular disease included type 2 diabetes mellitus and systemic hypertension diagnosed 1 and 2 years earlier, respectively, and 10 pack-years of cigarette smoking. There was no history of recurrent upper respiratory tract infection. He is married to 2 wives and has 12 children. His senior sister has also been on treatment for type 2 diabetes. Physical examination showed evidence of biventricular failure. He had raised blood pressure (140/100 mmHg) and features of long standing hypertension including thickened arterial wall, locomotor brachialis and grade II Keith-Wagener-Baker hypertensive retinopathy. The apex beat and heart sounds were on the right side of the chest. There was loss of sensation to light touch involving the feet and distal one quarter of both legs ("stocking distribution"). The body mass index was within normal range (20.0 Kg/m^2).

Chest radiograph (Figure 1) showed cardiomegaly (cardiothoracic index = 58.4%) with left ventricular preponderance and upper lobe vessel diversion. The cardiac apex, aortic arch and gastric bubble were on the right side of the chest. The left hemi-diaphragmatic dome was higher than the right by about 3.5 cm. Abdominal ultrasound showed that the liver and the spleen were normal but located in the left and right upper abdomen, respectively. Left sided electrocardiogram (ECG) (Figure 2a) demonstrated negative P wave deflection in leads I and aVL with R wave regression in the praecordial leads - mirror image of normal. Though the P waves were inverted in lead I and aVL of the right sided ECG, R wave progression in the praecordial leads was normal. The right sided ECG also show T wave inversion in leads V_3-V_6. Both tracings showed low QRS wave amplitude. Echocardiography done using right sided chest echo windows confirmed the location of the heart in the right hemithorax. Abnormal echocardiography findings included thickened left ventricular wall, ejection fraction of 30%, and mitral and tricuspid valve regurgitation. There were however neither co-existing congenital cardiac/great vessel anomaly nor abnormality of valve morphology, myocardial wall and segmental motion.

Computed tomogram scan also confirmed the location of the heart in the right hemithorax, and reversal of the normal positions of the abdominal viscera (stomach, liver, gall bladder and spleen) (Figure 3). These thoracic and abdominal organs were structurally normal. Apart from elevated fasting blood glucose (12.9 mol/L), urinalysis and blood chemistry including electrolytes, creatinine, lipid profiles and cardiac enzymes (Aspartate Transaminase and Myocardial Bound Creatinine Kinase) levels were normal.

A diagnosis of hypertensive diabetic heart disease with systolic dysfunction in a patient with dextrocardia and situs viscerum inversus totalis was made. He was counselled and advised to cease

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cigarette smoking. The heart failure responded satisfactorily to anti-heart failure medications including loop diuresis, angiotensin converting enzyme and aldosterone inhibition, and low dose beta blockade with carvidilol. Blood sugar was adequately controlled using metformin and glibenclamide. Blood pressure control was achieved without additional antihypertensive medication.

Figure 1: **Chest X-Ray Showing the Cardiac Apex Pointing Towards the Right Side of the Chest.**

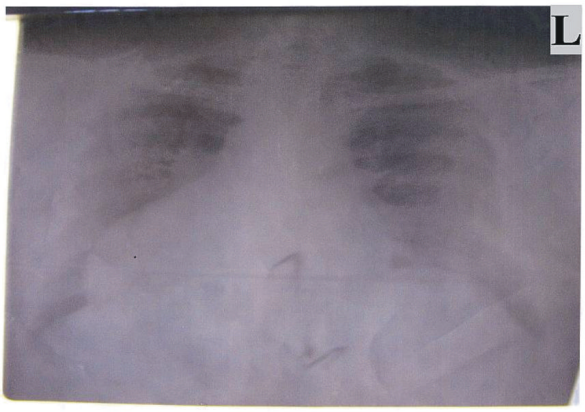


Figure 2: **(a) Left and (b) Right Sided Electrocardiography.**

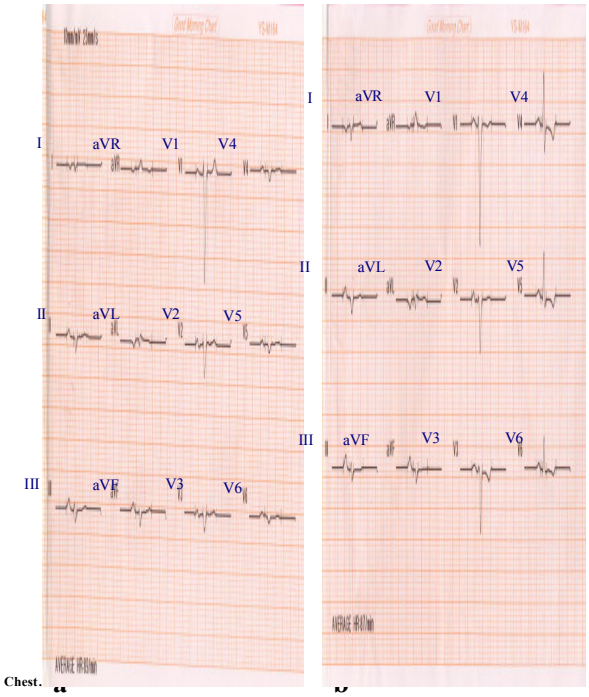
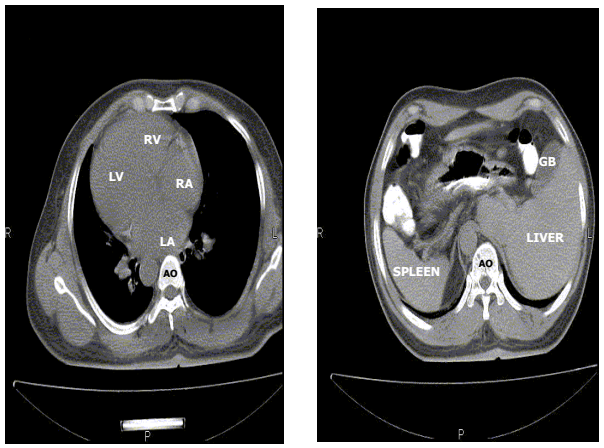


Figure 3: **Computed Tomogram Scan Showing (a) Dextrocardia and (b) Abdominal Visceral Situs Inversus.**



a **b**

LEGEND
AO: Aorta, GB: Gall bladder, LA: Left Atrium
LV : Left ventricle, RA: Right atrium
RV : Right ventricle

DISCUSSION

Dextrocardia is a rare developmental anomaly in which the primitive heart loop folds to the left in a mirror image of a normal bulboventricular loop. This result in the positioning of the heart in the right side of the thorax with the cardiac long axis directed to the right and inferiorly. Dextrocardia occurs in about I per 10, 000 in the general population [1,2]. This is lower than an incidence rate of 1 in 12,019 pregnancies reported in a retrospective hospital-based study involving 85 cases of dextrocardia [3]. Though the exact cause is unknown, dextrocardia has been linked with a number of factors including autosomal recessive gene with incomplete penetrance, maternal diabetes, cocaine use, and conjoined twinning [4-6]. In addition to being diabetic the current case report has family history of diabetes though this does not necessarily suggest a cause and effect relationship. The diagnosis of dextrocardia is made using combination of clinical, radiological, electrocardiography and echocardiography findings. Situs inversus dextrocardia may be diagnosed incidentally during routine medical examination or evaluation for unrelated conditions as in the current case report.

There are no data on the incidence of congenital cardiac malposition in Nigeria. Of the five cases of dextrocardia that has been documented, to the authors' knowledge, in Nigeria, two had no co-

Existing cardiac anomaly and were asymptomatic; while one and two of the remaining three had cardiac and noncardiac symptomatic malformations, respectively. [7-10]. Electrocardiography is an important and interesting tool in the diagnostic Armamentarium of dextrocardia. Reversed arm ECG may be misdiagnosed as dextrocardia because the QRS and P waves are negative in lead I in both conditions (Figure 2a and 2b). However, QRS wave progresses normally in the praecordial leads in reversed arm ECG (Figure 2b) while it regresses in dextrocardia (Figure 2a). Echocardiography is required for precise analysis of cardiac anatomy as well as detection of associated congenital cardiac or great vessel malformation. Sonographic and computed tomogram studies are required to confirm the anatomical positions of abdominal visceral.

The arrangements of the position of the abdominal viscera in dextrocardia may be normal (situs solitus), reversed (situs inversus), and indeterminate (situs ambiguous or isomerism) in 32-35%, 35-39% and 26-28% of cases, respectively [1-3]. Dextrocardia with a normal abdominal situs has a high incidence of associated congenital cardiac anomalies including among others, transposition of great vessels, and atrial and ventricular septal defects in 90-95% of cases. On the other hand, dextrocardia with situs inversus is associated with a lower incidence of congenital heart disease (0-10%) [1-3, 11,12.] Non-cardiac malformations, on the other hand, occur in the same proportion in situs inversus and ambiguous [1]. One such non-cardiac malformation is the combination of dextrocardia, abnormality of mucociliary function and bronchiectasis (Kartagener's syndrome). This rare triad has been reported in a Nigerian child [8] and may be associated with male infertility. About 46% and 48% of patients with primary ciliary dyskinesia may have situs solitus and inversus totalis respectively, [13,14]. The current case report had neither pulmonary nor reproductive abnormality and the observed concordant atrioventricular connection has been reported in 74.3% of patients with situs inversus dextrocardia [12, 13]. Patients with situs inversus dextrocardia are, like the general population, susceptible to acquired cardiac diseases. Though the current case report had classical risk factors of ischemic heart disease, there was no evidence of coronary artery lesion. Nonetheless coronary artery disease has been reported in patients with situs inversus dextrocardia [15,16.] The current case is to our knowledge the oldest reported in Nigeria, and illustrates that patients with situs inversus dextrocardia can have normal life expectancy like the general population.

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