

# Dextrocardia, Situs Inversus And Multiple Congenital Cardiac Defects In A Nigerian Infant

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## Abstract

Situs inversus with dextrocardia is the complete reversal of the position of the abdominal and thoracic viscera. It may remain asymptomatic or could be detected early in infancy when associated with other congenital malformations especially of the cardiovascular system. Situs inversus with dextrocardia usually exists without co-existing congenital heart disease. We report the case of a seven month old Nigerian male infant who presented with complaints of recurrent cough and bluish discoloration of the lips. Examination revealed central cyanosis with grade 3 digital clubbing and a right-sided apical impulse. Chest radiograph showed a right sided heart and Echocardiogram confirmed multiple cardiac anomalies (Complete Atrio-ventricular septal defect (AVSD), Pulmonary atresia and Patent ductus arteriosus).

## INTRODUCTION

Dextrocardia is a congenital anomaly in which the heart is situated on the right side of the body<sup>1</sup>. Situs inversus is a condition in which the major visceral organs are mirrored from their normal positions, that is the morphological left atrium is on the right while the morphological right atrium, the liver and gall bladder are on the left.<sup>2</sup> This is otherwise known as Situs inversus totalis or mirror image dextrocardia, which is a rare condition with a prevalence of 1:10,000 in some populations.<sup>3</sup> It is generally asymptomatic except when associated with congenital heart defects which is seen in 5 to 10% of cases.<sup>3</sup> Various cardiac anomalies have been described in literature including Ventricular septal defects<sup>5,6</sup> and Transposition of the great arteries<sup>6,7</sup>. Here we report a case of an infant presenting with dextrocardia, situs inversus and multiple congenital heart anomalies.

## CASE REPORT

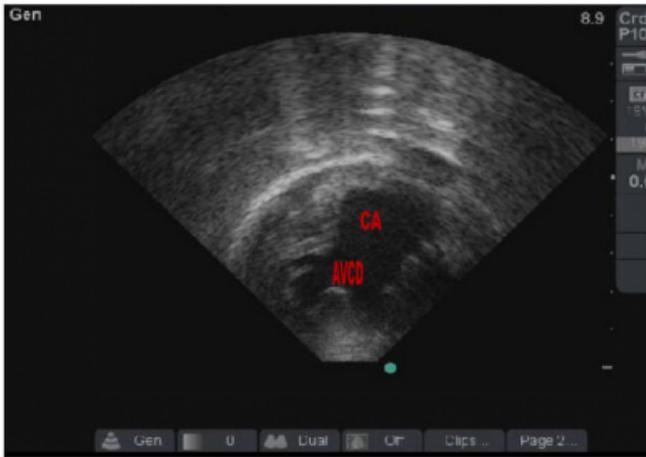
A seven month old male Nigerian infant presented at the Children outpatient clinic of the Niger Delta University Teaching Hospital Bayelsa with complaints of recurrent cough and bluish discoloration of the lips. On examination, he was found to be centrally cyanosed, Spo<sub>2</sub> (55%) with grade 3 digital clubbing and no dysmorphic features except for widely spaced nipples. His weight was 7.6kg and height was 62cm. His peripheral pulses were palpably normal. Cardiac apical impulse was located in the fourth right intercostal space. The second heart sound was single and soft

and there was no murmur. There was no evidence of cardiac failure.

He was the second of two children and the product of term gestation delivered by spontaneous vertex to a 32 year old mother in a non-consanguineous marriage. There was no history of exanthematous febrile illness, ingestion of herbal concoctions or exposure to irradiation in pregnancy. The labour and delivery were uneventful and there was no history of birth asphyxia. His birth weight was 3.2kg. The neonatal period was uneventful. There was no history of feeding difficulties but occasional respiratory distress. His packed cell volume (PCV) was 55% while the oxygen saturation ranged from 53-61% in room air. Chest radiograph revealed dextrocardia with situs inversus.

**Figure 1**

Fig 1 showing Common atrium (CA) and Atrio-ventricular canal defect (ACCD)



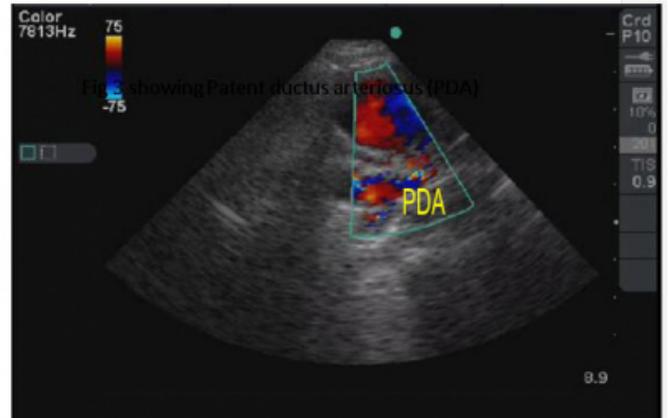
**Figure 2**

Fig 2 showing common atrio-ventricular valve (CA -Vv) and ventricular septal defect (VSD)



**Figure 3**

Fig 3 showing patent ductus arteriosus (PDA)



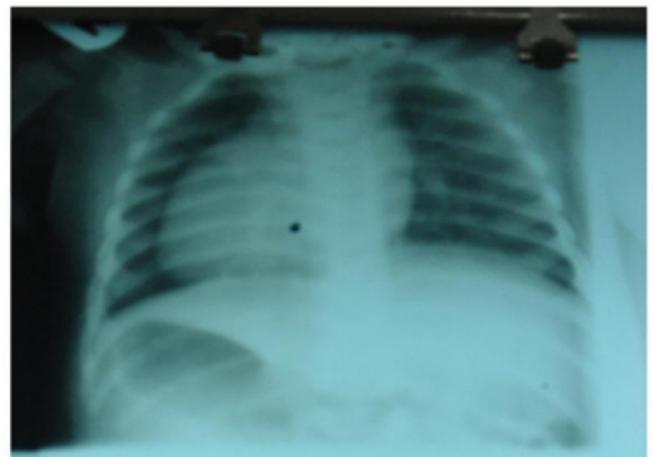
**Figure 4**

COMMON A-V VALVE REGURGITATION



**Figure 5**

Anteroposterior Chest radiograph showing the cardiac apex and gastric air bubble on the right



## **DISCUSSION**

Dextrocardia is a congenital defect in which the heart is situated on the right side of the body. It was first recognized in 1643 by Marco Severino.<sup>1</sup> Situs inversus is a congenital condition in which major visceral organs are mirrored from their normal positions, that is, the morphological left atrium is on the right while the morphological right atrium, the liver and gall bladder are on the left.<sup>2</sup> This was first described by Matthew Ballie more than a century later.<sup>1</sup> The association of situs inversus with dextrocardia is also known as Situs inversus totalis, which is a rare condition occurring in about 1:10,000 of the general population.<sup>4</sup> Situs inversus is generally an autosomal recessive genetic condition, although it can be x-linked or found in identical twins.<sup>1, 6</sup> Dextrocardia with situs inversus has been known to be associated with a lower incidence of congenital heart disease (0-10%) unlike its association with situs solitus (the normal laevoposition of the heart) with up to 90% of cases.<sup>8, 9, 17</sup> Common congenital cardiac defects reported include transposition of the great arteries<sup>6</sup> and ventricular septal defects<sup>8, 10</sup> among others.<sup>17</sup> Rarer cardiac anomalies include hypoplastic left heart syndrome which was reported in a cyanotic neonate at birth.<sup>3</sup> Patients with dextrocardia and situs inversus are usually asymptomatic and discovered usually during routine examination as has been found in many Nigerian studies.<sup>8, 11-14</sup> This was in contrast to our case where the child showed early symptoms of the underlying congenital heart disease, thus leading to the earlier presentation.

Extra-cardiac malformations have also been noted to be associated with Situs inversus and dextrocardia. A case of Katagener syndrome has been reported in a Nigerian child<sup>15</sup> with associated sinus and pulmonary infections. Eze et al<sup>12</sup> reported a case of a Nigerian infant with dextrocardia, undescended testes and polydactyl of the hands and feet. A review of the literature shows a case of multiple congenital anomalies: situs inversus totalis with atrioventricular septal defect, double outlet left ventricle, coarctation of the aorta as well as other extra cardiac malformation in a 23 week old foetus which were not compatible with life.<sup>17</sup> A case of a neonate who had situs inversus and dextrocardia in association with Goldenher syndrome was also reported.<sup>16</sup> This syndrome involved dysmorphogenesis of the first and second brachial arches and was associated with cardiac as well as vertebral and ocular anomalies. In our case, the presence of wide spaced nipples was the only dysmorphic feature in this child and there was no other suggestion of any other associated

syndrome.

## **CONCLUSION**

This case highlights the presence of atrioventricular septal defect with situs inversus and dextrocardia which has rarely been described in literature, in addition to the pulmonary atresia and patent ductus arteriosus. Dextrocardia with situs inversus is a congenital malformation which is rarely associated with cardiac defects and rarer still with A-V canal defects. The presence of this rare combination in an infant in a developing country with no Cardiac centre and high risk of mortality is highlighted.

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