*Nigerian Journal of Paediatrics* 2005; 32: (4) 94-96

# *Challenges in The Management of Congenital Heart Disease in Contemporary Nigeria: An Illustrative Case of Transposition of The Great Arteries*

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Summary

**Fadero FF, Oyedeji OA, Aremu AA, Ogunkunle OO. Challenges in The Management of Congenital Heart Disease in Contemporary Nigeria: An Illustrative Case of Transposition of The Great Arteries** *Nigerian Journal of Paediatrics* **2005; 32: 93.** The case of a three-month-old Nigerian girl with Transposition of the Great Arteries is presented. The definitive diagnosis was based on echocardiography. The parents were counselled on the need for better care in a paediatric cardiology unit where surgical intervention would be offered as the definitive management of the disease sometime in the near future. However, due to financial constraints, the parents failed to take the baby to the specified referral hospital weeks after the referral. This case illustrates the challenges in the management of congenital heart diseases in present day Nigeria as well as the need to establish cardiac centres in the country, where such cases would be properly managed.

## Introduction

TRANSPOSITION of the great arteries (TGA) is the commonest cyanotic congenital heart disease presenting in early neonatal period and constitutes 4.5-7 percent of all congenital heart diseases with a male preponderance.[1-3] In this condition, there is ventriculo-arterial discordance, with the aorta arising from the morphologic right ventricle and the pulmonary artery from the morphologic left ventricle leading to two parallel circulations.[1] There are two major anatomic forms of the lesion, the commoner form being the dextro-Transposition of the Great Arteries (d-TGA) where the aorta is anterior and to the right of the pulmonary artery; while the other is the laevo-Transposition of the Great Arteries (l-TGA) in which the aorta is to the left of the pulmonary artery.[1]

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This situation is incompatible with life, except there is a shunt such as a patent ductus arteriosus (PDA), atrial septal defect (ASD) or ventricular septal defect (VSD) linking the parallel circulations and allowing for mixing of venous and arterial blood. Where any of these shunts is non-existent, the administration of intravenous prostaglandin very early in the neonatal period as a medical palliative measure, is useful in sustaining ductal patency in the attempt to prolong life. In the event of failure of this palliative measure, a Balloon (Rashkind's) atrial septostomy is a useful alternative that will increase intracardiac mixing of blood. Surgical options for palliation or correction include Blalock-Hanlon atrial septectomy, Mustard's or Senning's operations as well as the Arterial Switch Operation.[4] Without any intervention whatsoever, half of the affected children die in the first month of life and 90 percent fail to witness their first bi thday.[1]

We present the case of a three-month old girl with TGA in order to highlight the challenges of management of the disease in present day Nigeria.

## Case report

A three-month-old girl presented to the Children's Emergency Unit of the Ladoke Akintola University of Technology Teaching Hospital, Osogbo, with a three-day history of inability to suck from the breasts, progressive breathlessness and fever. Two days earlier, the parents had noticed that her extremities were blue.

Her 39-year old mother, a petty trader in the local market in their home town which was a considerable distance from the Ladoke Akintola University Teaching Hospital, had been worried by her slow growth since birth when compared with her older siblings. She was a product of a full term uneventful pregnancy supervised in a private hospital. She was born spontaneously via the vaginal route and cried immediately after birth. Her birth weight was 3,650 grams and she was discharged home on the day of delivery. She was noticed to be jaundiced at the age of three days but this cleared at the end of the first week of life without any significant treatment. Her 45-year-old father worked as a civil servant with the Local Government Authority in their hometown. All her four older siblings are alive and well. No family history of congenital heart disease or any other malformations had been reported in any of the family members.

General physical examination revealed an acutely ill and breathless infant with central cyanosis and a temperature of 38.2°C She was small for age, her weight being 3,800 gm. She had no dysmorphic features, but there was an obvious bulge over the praecordium. Her pulse rate was 160 beats per minute, full and regular. The cardiac apex was located at the 5th left intercostal space along the mid-clavicular line. Both first and second heart sounds were normal but there was a grade 4/6 continuous murmur heard loudest in the pulmonary area. The respiratory rate was 68 cycles per minute. The breath sounds were vesicular and there were no added sounds. The abdomen was soft and the liver was palpable 4cm below the right costal margin; it was firm and non-tender. No other masses were palpable in the abdomen. All the other systems were essentially normal.

The initial assessment was that of a cyanotic congenital heart disease with probable patent ductus arteriosus, in heart failure. Not all the investigations requested were carried out because of parental financial constraints. However, the results of those done at various times in the course of her admission were as follows: packed cell volume (PCV) = 48 percent; a blood film examination showed 2+ of P. falciparum; the blood culture did not yield any organism; a chest radiograph (Fig 1) showed a cardio-thoracic ratio of 0.78 and the classical "egg-on-side" appearance often found in TGA; the lung fields were clear. The electrocardiograph (ECG) showed sinus tachycardia, right axis deviation (+127°) and both right atrial and right ventricular hypertrophy. Two-dimensional (2-D) echocardiography revealed situs solitus, atrio-ventricular concordance and ventriculo-arterial discordance; the anatomic right and left ventricles were both hypertrophied; the aortic root

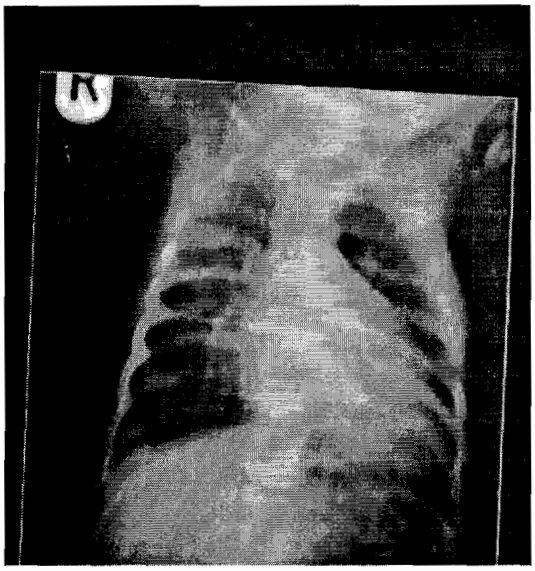

Fig 1: *Chest X-Ray showing the classical "egg-on-side" appearance in TGA*

was anterior and parallel to the pulmonary root; there was a PDA but no VSD or ASD was demonstrated. Thus, the overall echocardiographic picture was in keeping with a d-TGA with a PDA.

The initial treatment included intranasal oxygen, digitalization and fluid administration at two-thirds of maintenance requirement. Intramuscular artemether as well as intravenous cefotaxime and gentamicin were also commenced. Breast-feeding was suspended, and feeding by nasogastric tube instituted. By the 5th day of admission, her fever had subsided, the heart rate ranged between 100–120 beats per minute and her liver had shrunken by 2cm. However, her respiratory rate ranged between 40 and 50 cycles per minute. She was considered well enough to resume breast-feeding. It was at this point that she had the echocardiography which revealed the morphology of her cardiac lesion.

On reviewing the echocardiographic findings and considering her clinical status, it was decided to refer her to the University College Hospital (UCH), Ibadan for further investigation and possible surgical intervention. Since it was the experience at our centre that none of the children with congenital heart diseases that had hitherto been referred by our Unit to UCH on outpatient basis, actually ever reported there, it was decided that this particular patient should be referred as an in-patient of the Unit. Following arrangement for immediate admission on arrival, the patient was referred to a paediatric cardiologist at UCH, Ibadan, a centre with better facilities, more experienced personnel and the possibility of surgical intervention. After a protracted course of counselling on the consequences of uncorrected TGA, the parents agreed to the referral but only reluctantly, on account of financial constraints. Unfortunately, many

weeks after the referral, the patient is yet to show up at the UCH and the parents are yet to communicate again with the doctors at the Ladoke Akintola University of Technology Teaching Hospital.

## Discussion

The incidence of congenital heart diseases in developing countries has been estimated to be between 6 and 8 /1000 live births and a previous study conducted among Nigerians with congenital heart diseases has reported a prevalence of 4.5 percent for TGA.[3] This makes TGA a relatively uncommon disease in clinical practice. Clinically, uncommon diseases are prone to being missed as was the case in this infant whose TGA was only diagnosed at the age of three months. Another factor responsible for a delay in an earlier diagnosis, particularly in the neonatal period, could be the fact that the disease may be asymptomatic especially where the degree of oxygenation through the existing PDA (inter-circulatory mixing) is high.[1] Perhaps if this child had been examined soon after birth, the murmur of PDA detected on admission could have alerted the examining clinician earlier to the possibility of a congenital heart defect. Such lapses as were responsible for the delayed diagnosis in this patient could lead to early deaths in cases of TGA.

The role of echocardiography in the diagnosis of TGA and other structural heart defects cannot be overemphasized, as previous studies have shown that this procedure is particularly useful in the diagnosis of structural heart problems.[5,6] Unfortunately, only very few hospitals in Nigeria can lay claim to echocardiography and the required skill. Cardiac catheterization, a highly invasive procedure that can also be helpful both in the diagnosis and palliative management of TGA, is now mainly useful where echocardiography has not completely demonstrated the anatomical lesions of the malformation.[1] To the best of our knowledge, there is no centre in Nigeria performing cardiac catheterisation at the present time.

In developed countries, advances in medicine have made the diagnosis and correction of TGA possible thereby increasing the life span of surgically corrected patients.[7] This is contrary to the situation in Nigeria and many developing countries where the poor state of health care delivery, low level of medical technology and health financing are exemplified in this patient. Standard care dictates that diagnosis in this patient could have been made even in utero[8] or in the early neonatal period by routine paediatric examination of the newborn. When she was referred to the UCH, the minimum acceptable means of transportation should have been an ambulance with a specialized medical team. The financial burden of managing this child rested squarely on the parents who were reluctant to take her to UCH mainly because of their poor financial status. It is hoped that as the recently established National Health Insurance Scheme in the country matures, the scheme will extend its scope to involve the management of patients with lesions of this nature so that the financial burden on the family would be less. It is also hoped that this scheme will reduce to a minimum, the rate of follow-up defaults, discharges against medical advice and the length of time wasted in counselling of parents with obviously very sick children about the need for medical attention for their wards.

In order to optimise the care of patients with any form of heart lesion, whether congenital or acquired, cardiac centres should be established in Nigeria, with manpower and equipment adequate for the teeming number of children with such lesions. Such centres could be a joint venture between the public and private sectors of the economy.

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