Case report of challenges in the management of a rare ductal dependent complex congenital heart disease in a Nigerian tertiary hospital

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Case Report

Abstract:
Objective: Complex congenital heart defects are rare and may be difficult to define. They often require early surgery for palliation or correction. A lack of facilities and manpower to provide surgery in developing countries often results in mortality.

Case report: A 6 month old male infant referred to our unit on account of failure to thrive, cyanosis since birth, easy fatigability and breathlessness. On examination he was small for age with tachycardia and a grade III pansystolic murmur at the left lower sternal edge. Chest radiograph revealed an "egg on side" cardiac appearance with cardiomegaly. Echocardiography confirmed the presence of d-transposition of the great arteries with a patent foramen ovale, large ventricular septal defect and atresia of the proximal main pulmonary artery. The child died while being prepared for referral to a centre for palliative surgery.

Conclusion: Transposition of the great arteries with pulmonary atresia is an uncommon congenital heart disease. Early intervention with palliative surgery is necessary to prevent mortality.

Keywords: Congenital heart disease; pulmonary atresia with ventricular septal defect; great vessel anomaly; echocardiography

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Rapport de cas de défis dans la gestion d'une maladie dépendante canalaire rare complexe cardiaque congénitale dans un hôpital tertiaire nigérian

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Rapport de cas

Résumé

Objectif: Complexe congénitales malformations cardiaques sont rares et peuvent être difficiles à définir. Ils nécessitent souvent une intervention chirurgicale précoce pour la palliation ou la correction. Un manque d'équipements et de la main-d'œuvre pour fournir une intervention chirurgicale dans les pays en développement se traduit souvent par la mortalité.

Rapport de cas: A 6 mois, vieux nourrisson de sexe masculin visé à notre unité en raison d'un retard de croissance, cyanose depuis la naissance, fatigabilité et dyspnée facile. À l'examen, il était petit pour l'âge avec tachycardie et un murmure holosystolique grade III sur le bord inférieur gauche du sternum. La radiographie thoracique a révélé un "oeuf sur le côté" apparence cardiaque avec cardiomégalie. L'échocardiographie a confirmé la présence de d-transposition des grandes artères avec un foramen ovale de brevet, un grand défaut du septum ventriculaire et l'atrésie de l'artère pulmonaire principale proximale. L'enfant est mort tout en étant préparé pour l'orientation vers un centre de chirurgie palliative.

Conclusion: Transposition des grandes artères avec atrésie pulmonaire est une maladie cardiaque congénitale rare. Une intervention précoce avec la chirurgie palliative est nécessaire pour prévenir la mortalité.

Mots-clés: maladie cardiaque congénitale; atrésie pulmonaire avec ventricular septal defect; grand navire anomaly; échocardiographie

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INTRODUCTION

Transposition of the great arteries (TGA) is an uncommon congenital heart disease with large series reporting an incidence of 20 – 30 per 100,000 live births (1,2). It is characterized by parallel pulmonary and systemic circulation with survival dependent on intermixing through shunts/ defects (1,3). The commonest variation has ventriculoarterial discordance with a ventricular septal defect(VSD) and clinical presentation occurring typically in the neonatal period with heart failure (1,3). Unusual variations of this condition have in addition, aortic abnormalities, left ventricular outflow tract obstruction overriding of the pulmonary artery and anomalous pulmonary venous return (1,3). Limited capacity for diagnosis, associated fatality rates and the relative rarity of these conditions have meant such complex congenital heart diseases have often gone unrecognised in reports from developing countries (4,5). Mortality in developing countries is often a consequence of the severity of the condition and delays in instituting appropriate corrective/ palliative interventions (5).

We present an unusual variation of transposition of the great arteries associated with pulmonary atresia in a Nigerian infant.

CASE REPORT

A six month old male infant presented to our unit with poor weight gain since birth and worsening breathlessness of two months prior to presentation. He had been fed with breast milk, water and herbal medications since birth. Breastfeeding was about eight times in a typical 24 hour period with duration lasting about 10-15min. Breathlessness was noticed about two months prior to presentation, present at rest and worsened by exertion. There was associated cyanosis which worsened in the last two months. Child had occasional cough and a history of excessive sweating while feeding. He was a product of a term pregnancy to a 27 year old mother, booked at 4 months gestational age. There was no maternal rash, febrile illness or diabetes in the mother during pregnancy. Child suffered perinatal asphyxia at delivery. Motor developmental milestones had been appropriate for his age. There was no family history suggestive a congenital heart disease.

On examination, he weighed 5 kilograms and had a length of 61cm. He was in severe respiratory distress, plethoric, centrally cyanosed, had grade III digital clubbing and no edema. Axillary temperature was 36.8^{0}C. He was tachypneic with a respiratory rate of 59breath/minute. Breath sounds were vesicular. Pulses were regular and full volume with no radiofemoral delay. No precordial hyperactivity but slight precordial bulge. Apex beat was situated at the 4th intercostal space anterior axillary line. His heart rate was 152 beats/minute with first, second heart sounds and a grade 3/6 pansystolic murmur heard loudest at the left lower sternal border. Hemoglobin oxygen saturation remained less than 70% on 100% oxygen. Other examination findings were within normal limits. A diagnosis of complex cyanotic congenital heart disease was made and he was commenced on 100% oxygen, parenteral morphine and received 1meq/kg of sodium bicarbonate slowly for acidosis. Complete blood count revealed a packed cell volume of 56%, hemoglobin of 12.1g/dl, red blood cell count of 7.1 x 10^{12} cells/L, mean corpuscular volume of 71fl, Mean corpuscular hemoglobin concentration of 24g/L, platelet count of 179 x 10^{9} cells/L and white blood cell count of 12.2 x 10^{9} cells/L with 78% neutrophils and 16% lymphocytes. Electrolytes, urea and creatinine were normal.

Chest radiograph (Figure 1) showed increased cardio-thoracic ratio of 8.4/11.5 with normal pulmonary vascularity. The cardiac contour was in keeping with an “egg on side” appearance. Patchy opacities were seen in both lung fields suggesting pneumonia.

Electrocardiogram showed a normal sinus rhythm. There was rightward deviation of the QRS axis (+165^{o}) with Tall R waves in V3R, deep S waves in V5 and V6, upright T waves inV3R and V1 and P wave amplitude in lead II of 3.5mm suggestive of right ventricular hypertrophy and right atrial enlargement. Echocardiography showed cardiac situs was solitus, atrioventricular concordance and discordant ventriculoarterial connections (Figure 2). The inferior vena cava was dilated with absent inspiratory collapse. The right ventricle was dilated and hypertrophied while the left ventricle was reduced in size, however not hypoplastic. The aorta was located anteriorly and rightwards arising from the right ventricle with a diameter of 1.99cm (Figure 2). The pulmonary annulus (located posteriorly and to the left arising from the left ventricle) and the proximal main pulmonary artery were atretic (Figure 2). The distal portion of the main pulmonary artery measured 0.89cm in diameter. There was a patent foramen ovale (PFO) measuring 0.29cm (Figure 3) and a large outlet ventricular septal defect

measuring 1.5cm. The ductus arteriosus was patent with a diameter of 0.29cm.The pulmonary veins were normally sited. There was bidirectional flow across the PFO and left to right flow across the VSD. There was aortic to pulmonary blood flow across the ductus arteriosus. A final diagnosis was made of complex congenital heart disease with D-transposition of the great arteries, pulmonary valvular atresia, patent foramen ovale, outlet VSD and patent ductus arteriosus(PDA). Facilities and manpower for performing palliative heart surgery were not available in the hospital. Attempts at maintain patency of the ductus arteriosus with prostaglandin infusion were unsuccessful as it was not available in the hospital and other tertiary hospitals in the region. While being planned for referral for palliative surgery, child developed increasing respiratory distress, began gasping and subsequently died at about 72 hours into admission. Parents declined our request for an autopsy for traditional/ cultural reasons.

DISCUSSION

In the largest study on congenital heart disease in Nigeria by Sadoh *et al*,(6) transposition of the great arteries constituted 1.5% of all congenital heart disease. Data from other countries report a rate of 5-7% of all congenital cardiac defects (3). The subset described in this report, transposition of the great arteries with pulmonary atresia is very rare and seen in 5-8% of all neonatal TGA (3). To the best of the author's knowledge, this is the first report of this rare subtype of TGA in Nigeria.

It is hypothesized that TGA arises from abnormal growth and development of the subaortic infundibulum and the absence of growth of the subpulmonary infundibulum resulting in the aortic valve being placed anteriorly compared to the pulmonary valve (3). Left ventricular outflow tract obstruction, when coexisting with a large VSD, is a consequence of anterior (leftward) displacement of the infundibular septum resulting in progressive obstruction and in severe cases atresia, as was demonstrated in the current case (3). Clinical presentation of this subset of presentations is similar to a severe form of Tetralogy of Fallot (TOF), however, a defining difference is that features in TGA with pulmonary atresia develop within the early neonatal period in contrast to TOF where cyanosis typically develops later (3). Double outlet right ventricle (DORV), previously described as a form of TGA may present in a similar way to TGA and, indeed, is a differential of the presentation of our patient. However, to be considered as DORV, it must be demonstrated that the pulmonary artery is also committed to the right ventricle which was not the case in our patient.

Children with D- TGA are dependent on intracardiac and extracardiac shunts for survival (1,3). Our patient had a PFO, large VSD and a patent ductus arteriosus that had maintained some degree of intermixing of the parallel circulations of TGA. The VSD was in this case largely responsible for maintaining pulmonary to systemic circulation flow while the PFO and PDA contributed to the systemic to pulmonary blood mixing. However, demands of growth and reduction of size of these defects/ shunts have been implicated in causing deterioration of the condition that may occur in late infancy (1,3).
Patients with TGA and pulmonary atresia and severely restricted pulmonary blood flow are recommended to have some form of palliative systemic to pulmonary arterial shunt early on, if corrective surgery is not feasible (1,3). Unfortunately, facilities and manpower to conduct these operations in small infants are unavailable in most parts of Nigeria. There exists an urgent need for cardiac surgical centers in Nigeria to commence surgical interventions for these complex disorders to avoid mortality.

CONCLUSION

This is an unusual occurrence of transposition of the great arteries with large VSD and pulmonary atresia. Delays in operative correction/ palliation may result in mortality. There is a need to develop capacity for the recognition and management of this condition in Nigeria.

Conflict of interest: The authors declare no conflict of interest.

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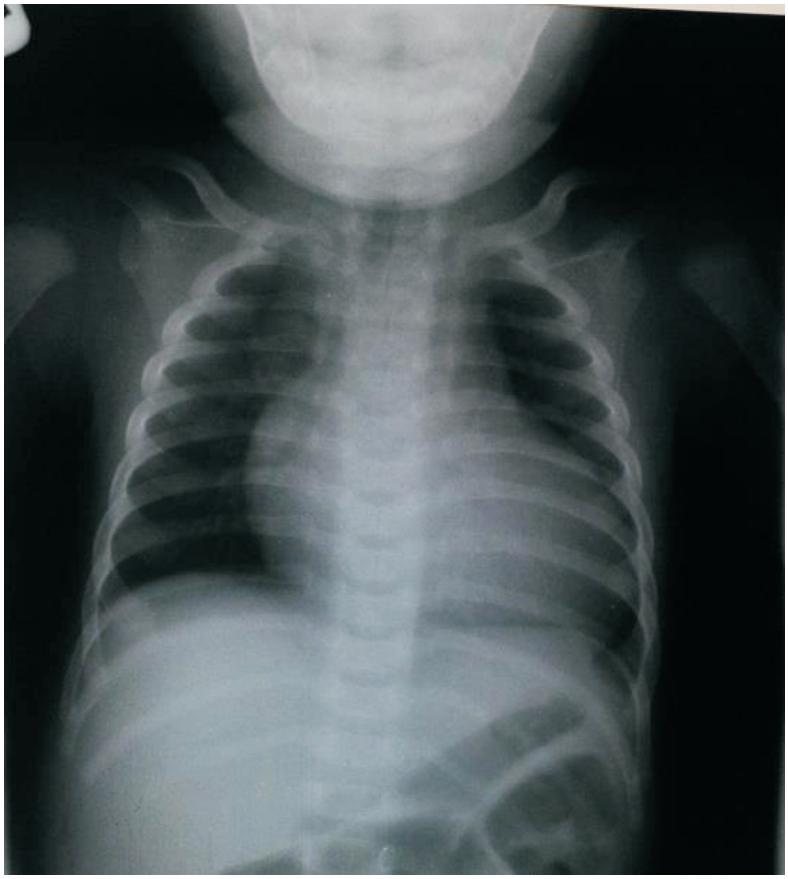

Figure 1: Chest radiograph depicting cardiomegaly with "egg on side" appearance

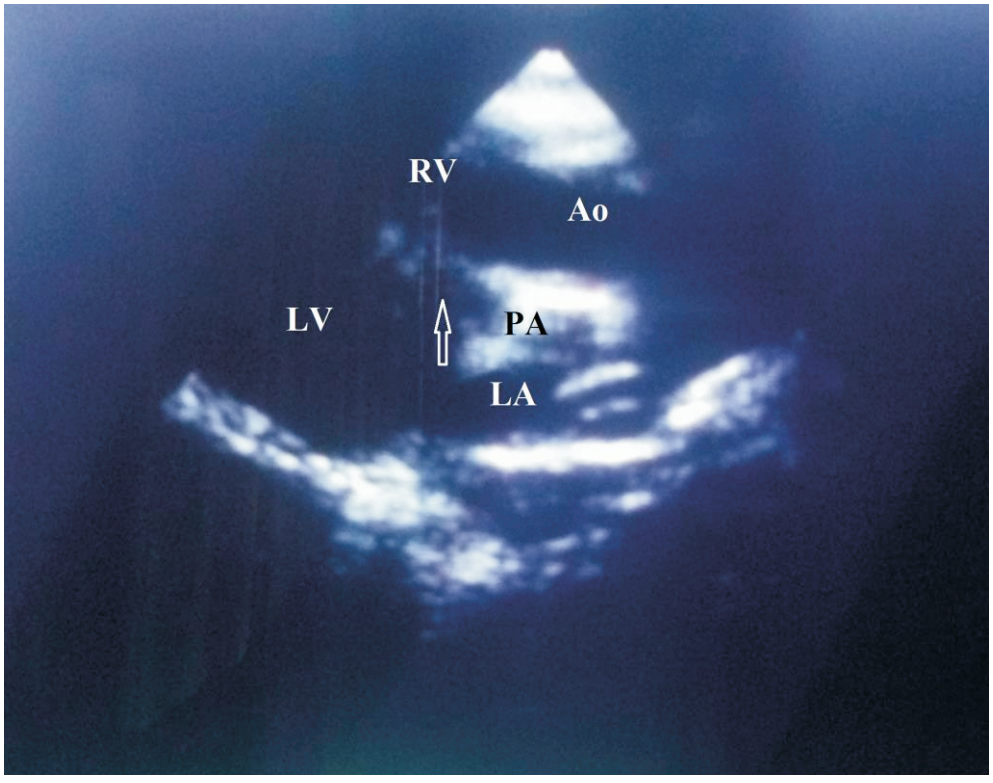

Figure 2: Parasternal long axis view showing the transposed great arteries, atretic proximal pulmonary artery and ventricular septal defect (arrow). Ao-Aorta, LA- Left atrium, LV-Left ventricle, PA- Pulmonary artery, RV-Right ventricle

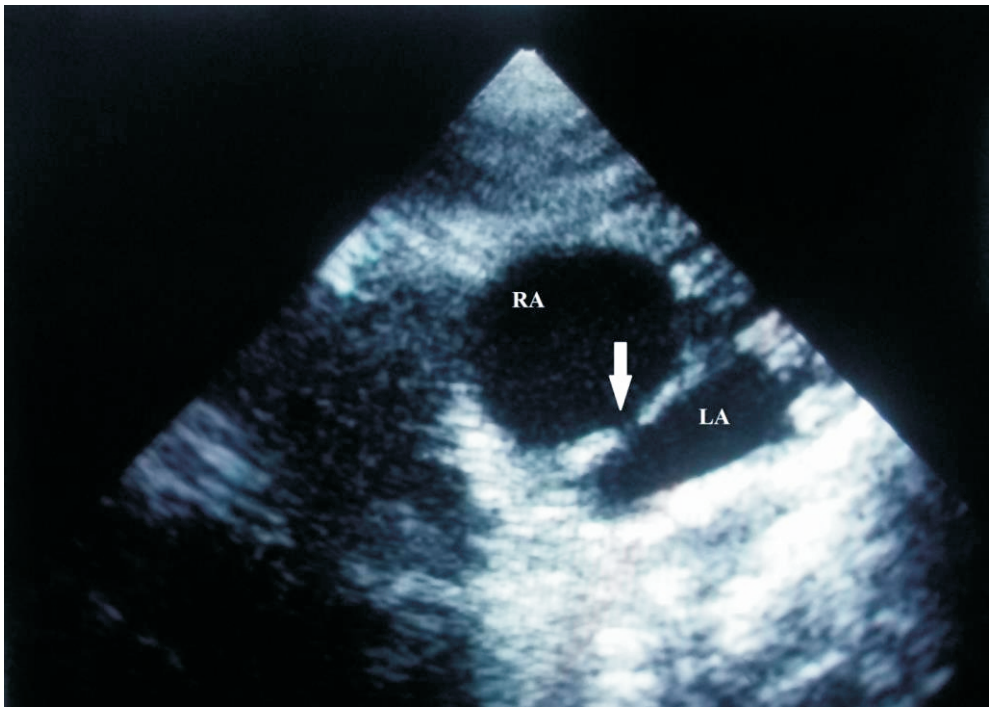

Figure 3: Subcostal atrial view showing the dilated right atrium (RA), smaller left atrium (LA) and patent foramen ovale (arrow)