Repair of Truncus Arteriosus, Type 1 in Nigeria: A Case Report

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Abstract

Truncus arteriosus, also known as common arterial trunk is an unusual embryologic anomaly. It is characterized by incomplete conotruncal septation resulting in a common aortopulmonary trunk and ventricular septal defect. The first surgical repair was performed in 1962 when Behndrett et al. closed the VSD and used a valveless conduit to establish right ventricle-pulmonary artery continuity. Complete repair of this anomaly with a valved conduit was first reported in 1967 by McGoon et al. and remains the procedure of choice for these patients. National Cardiothoracic Center of Excellence was established in 1984. Since then it had carried out significant surgical management of simple CHD. Recently, with the aid of foreign medical missions, it was able to carry out a successful repair of truncus arteriosus type 1, the first of its kind in Nigeria.

Keywords: Truncus arteriosus; Aortopulmonary trunk; Congenital heart disease; Hyperreflexia; Hydronephrosis


Introduction

Nigeria, a coastal West African country, has a population of about 170 million people. It has no fully established functional cardiology center for surgical management of adult and congenital heart diseases [1]. Currently, such cases are managed on adhoc basis (medical missions from foundations based in USA, UK and India). Such centers providing such medical services in Nigeria include University of Nigeria Teaching Hospital, UNTH, Enugu; Lagos University Teaching Hospital, LUTH; Lagos State University Teaching Hospital, LASUTH and Gwagwalada University Teaching Hospital, Abuja. Recently, in one of such missions at UNTH, Enugu, a complex and rare congenital heart disease, truncus arteriosus, type 1 was found and successfully managed, hence this report.

Truncus arteriosus is an uncommon congenital heart disease, accounting for less than 3% of all congenital heart diseases [2-4]. The features of truncus arteriosus are a single great artery arising from the base of the heart that supplies systemic, coronary and pulmonary blood flow, and ventricular septal defect. So many classifications exist for this rare defect but the most and clinical useful ones are those by Collet and Edward in 1949 and Van Praagh in 1965 [5]. The natural history is 80% mortality in the first year of life especially during infancy [6]. The result of physiological repair has remarkably improved over the years but pulmonary hypertension in the immediate postoperative period continues to pose a challenge. Historically, truncus arteriosus repair was first performed by McGoon et al. [7], with the use of aortic homograft including the aortic valve, with the aim of establishing continuity between right ventricle and pulmonary artery. From then till now, various methods have been developed with confusion over which method is best suited vis-a-vis the age at presentation. The frequently observed complications include conduit stenosis, truncal valve regurgitation or incompetence and or stenosis. High mortality and morbidity attend reoperation to address the listed complications.

This case report is aimed at stating unequivocally that truncus arteriosus among other complex congenital heart diseases are present in Nigeria and that a cardiac center, at University of Nigeria Teaching Hospital, Enugu has well equipped intensive care unit and personnel to manage the intricate postoperative challenges associated with its repair. However, trained local paediatric surgeon(s) are lacking, hence the foreign medical missions.

Case Report

OD is a 15 month male who lives with the parents at No 13B Jaja street, Aba, Abia state. Nigeria. They are Christians of the Redeemed Christian Mission. He presented at our service, within 4 months illness, at the age of 11 months with complaints of fast breathing that was noticed at 10 months of age, poor weight gain noticed at 9 months and 2 weeks, stiffness of the body noticed at 8 months, bluish discoloration...
of the lips, inability to pass urine and fever noticed, all prior to presentation 4 months ago. For the above complaints, he was initially managed at another hospital. There, clinical and echocardiographic diagnosis of d-TGA, large sub-aortic VSD including moderate aortic regurgitation was made. He was subsequently referred to our center for expert management.

At our center further evaluation showed that he was acutely ill looking male in respiratory distress (RR = 60 breaths/min), tachycardia (pulse rate = 160 beats/min), deep cyanosis (SPO$_2$ = 48% on room air), febrile (temp = 38°C), dry mucosa, nil peripheral oedema and no lymphadenopathy. His weight was 6 kg (60% of expected weight), length = 56 cm (low for age) and OFC= 41 cm (88.2% of expected). He had precordial bulge with displaced apex beat to 5th LICS, lateral to mid clavicular line. Heart sounds 1 and 11 with grade 3/6 pansystolic murmur maximal at the lower left sterna border with loud P2.

Liver was enlarged 8 cm below the right costal margin, soft and tender. Both kidneys were ballotable. Lung fields were clear. He was conscious but irritable, normal fontanel, in episphontic posture with global hypertonpia and hyperreflexia. The working diagnoses of CCF from cyanotic CHD, cerebral palsy from perinatal asphyxia, bilateral hydrenephrosis, with moderate hydration were made.

He was admitted into the ward and catheterized. Abdominal ultrasound done later excluded Hydrenephrosis and urine output improved on hydration and he was subsequently managed in collaboration with the pediatric neurologists, dieticians, physiotherapists and health visiting unit. He was discharged after 7 days to continue multidisciplinary follow up.

He was subsequently followed up awaiting surgical management. However, during one of the surgical cardiac missions at our center led by Dr Williams Novick cardiac Alliance, patient was re-evaluated for surgical management. During then, a repeat echocardiography was done and the following findings were noted: single outflow tract becoming aorta and pulmonary arteries, large VSD, moderate truncal valve regurgitation; RPA = 8.0 mm, LPA = 7.0 mm and coronary sinus was dilated. The diagnosis was then changed from d-TGA to truncus arteriosus type 1.

He was worked up for surgery with the results of the following investigations satisfactorily obtained. They included HBG, ESR, LFT, HBsAg, HCV, HIV 1 and 11, SEUC, Urinalysis, with urine MCS, PT/INR and C x R. Others were 12 lead ECG, ASO titre. Thereafter, 3 units of packed cells were cross-matched for him. Also platelets concentrates and FFP were provided. Informed consent of his parents was obtained.

Patient had open heart surgery for intra-cardiac repair. Patient was positioned supine, with GA with ETT and muscle relaxation with full cardiopulmonary monitoring. Cleansing and draping for standard full cardiopulmonary bypass. No pericardial or pleural effusion. He was weaned off ventilator and extubated successfully on the 14th post-operative day. He was placed on T-piece and continued to saturate at 95-96% on 2 L/min intranasal oxygen and 90% on room air (Figure 3). He had prolonged hospital stay due to unstable sternum. Sternal stabilization was done on the 32nd DPO. 11 days later he was discharged, precisely on the 43rd DPO in optimal condition. With RR = 28-36 breaths/min, saturation = 100% on room air, pulse rate = 108 beats/min, regular and moderate volume and BP = 114/60 mmHg. Current drugs at the time of discharge were oral frusemide, spironolactone, enalapril, cefixime and loratidine. He has been followed up at out-patient clinic on 2-weekly basis for more the 6 months with uneventful condition (Figure 4).

**Discussion**

Truncus arteriosus is an uncommon lesion. It accounts for 0.4-3.9% of autopsy proven cases of congenital heart defects [8]. As has been previously described, the lesion is characterized by single large arterial trunk originating from the base of the heart directly superior to a high VSD. PA originates from the trunk [9]. The defect is now correctable by closing the VSD and inserting a conduit between the right ventricle and pulmonary artery [10]. In the index patient, the conduit was a tabularized pericardium with a monocusp valve. The truncal valve which was competent was assigned to the aorta.

![Figure 1: Sternotomy done with native pericardium being harvested.](image1.jpg)

![Figure 2: Patient under full cardiopulmonary bypass with VSD being repaired.](image2.jpg)
Early repair has a better outcome because delaying the repair beyond 100 days carries the risk of postoperative pulmonary hypertensive crisis and cardiac failure [17,18]. Our patient was 15 months of age.

Our patient is still being followed up and as at the time of this report, no known late postoperative complications like recurrent supraventricular tachyarrhythmias, subacute bacteraemic endocarditis and right ventricular dysfunction have been observed.

**Conclusion**

Congenital heart diseases, both simple and complex, are present in our country. Making accurate diagnosis can be quite challenging unlike the developed world. Also important is the issue of repairing the complex types because of the dearth of requisite personnel as well as the equipment. Foreign cardiac missions that are humanitarian in nature can bridge the gap if consistent.

**References**