International Journal of Cardiovascular and Thoracic Surgery
2022; 8(3): 24-28
http://www.sciencepublishinggroup.com/j/ijcts
doi: 10.11648/j.ijcts.20220803.11
ISSN: 2575-4866 (Print); ISSN: 2575-4882 (Online)

Pattern of Truncus Arteriosus in Obafemi Awolowo University Teaching Hospitals Complex, Ile-Ife, South West Nigeria

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To cite this article:

Olukemi Bamigboye-Taiwo, John Akintunde Okeniyi, Babajide Samson Adeyefa, Olugbenga Olalekan Ojo, Uvie Ufuoma Onakpoya, Joel Oghenevware Eyekpegha, Abayomi Oguns. Pattern of Truncus Arteriosus in Obafemi Awolowo University Teaching Hospitals Complex, Ile-Ife, South West Nigeria. *International Journal of Cardiovascular and Thoracic Surgery.* Vol. 8, No. 3, 2022, pp. 24-28.
doi: 10.11648/j.ijcts.20220803.11

Received: March 29, 2022; **Accepted**: April 25, 2022; **Published**: May 24, 2022

Abstract: Background: Truncus arteriosus (TA) is a rare complex cyanotic congenital heart disease (CCHD) with significant morbidity and mortality. There are few reports on the pattern of TA in Nigeria. Objectives: To determine the pattern of TA in patients at the Obafemi Awolowo University Teaching Hospitals Complex, Ile-Ife, Nigeria. Methods: A three-year retrospective audit of the hospital records including echocardiograms and cardiac tomography scans where available, of consecutive patients diagnosed with TA was undertaken. Results: Twenty children, 10 boys and 10 girls had TA; median age at diagnosis was 6.5 months, range: 11 days-90 months, mean (SD): 16.8 ± 21.8 months. The mean weight was 7.3 ± 4.4 kg, range: 2.65-17.0 kg and the mean oxygen saturation was 72.5 ± 17.3%, range: 36-89%. Sixteen children (80.0%) had type I and 4 (20.0%) had type II TA. Types III and IV TA were not seen. The truncal valve was tricuspid in 12 (60.0%), quadricuspid in 7 (35.0%) patients, and bicuspid in one (5.0%) patient. Truncal valve regurgitation occurred in 10 (50.0%) patients; 60.0% were mild. Associated cardiac defects included patent ductus arteriosus [PDA (15.0%)], mitral atresia (15%), atrial septal defects, major aortopulmonary collateral arteries (10%), left superior vena cava (5%), tricuspid atresia (5%), common atrium (5%). Conclusions: Although TA is a critical CCHD, diagnosis is often late in our setting. Type I is the most common variant and associated anomalies include PDA, mitral and tricuspid atresia.

Keywords: Truncus Arteriosus, Pattern, Echocardiogram, Associated Anomalies

1. Introduction

Congenital heart disease (CHD) is a rapidly increasing global problem in child health with about 90% of affected children living in parts of the world with little or no facilities for care and thus mortality remains high [1]. Although there has been significant improvement in the diagnoses and treatment of CHD over the past eighty years, these successes are not recorded in the developing world where a significant number of deaths from CHD still occur [1]. Truncus arteriosus is a relatively rare complex cyanotic congenital heart disease representing about 2-4% of all congenital heart anomalies [2-4]. Truncus arteriosus (TA) is associated with significant morbidity and mortality [4]. The exact prevalence of truncus arteriosus in Nigeria is largely unknown because few reports have documented the disease prevalence in the country.

The anatomy of truncus arteriosus involves a single arterial trunk arising from a common ventriculoarterial junction and supplying the coronary, systemic, and pulmonary pathways [4]. The ventricular septal defect in truncus arteriosus is often large and occurs because of the absence of the infundibular septum [5]. The single semilunar valve in TA differentiates it from aortic and pulmonary valve atresia. In these conditions, a single arterial vessel similarly receives the entire output of both ventricles, however, a second atretic semilunar valve is present [5]. Two major classification systems describe the anatomy of TA: the Collett and Edwards [6] and Van Praagh classifications [7].

According to Collett and Edwards, [6] in Type I, a short pulmonary trunk originates from the truncus arteriosus and gives rise to two branch pulmonary arteries. In type II, each pulmonary artery arises independently from the truncus arteriosus close to one another, while in type III, they are at some distance from one another. In both types II and III, the main pulmonary artery is non-existent. Type IV is associated with underdevelopment of the aortic arch, including tubular hypoplasia, discrete coarctation, or complete interruption.

The clinical manifestation in TA is based on the anatomical type and other associated malformations. Majority of patients present with cyanosis and tachypnea, which may not be detected in the immediate neonatal period [8]. Diagnosis and detailed anatomical description of the lesion can be adequately made in most patients with transthoracic echocardiography. Other imaging modalities such as cardiac computed tomography angiography, or cardiac magnetic resonance imaging and cardiac catheterization may be indicated in those with more complex anatomy [8].

The definitive management of TA is total repair in the neonatal period [8]. This involves resection of branch pulmonary arteries from the arterial trunk with the placement of a right ventricle-to-pulmonary artery conduit and closure of the ventricular septal defect with a patch. In most established centres surgical outcomes are excellent. It is expected that over time the child will outgrow the conduit and would need reoperation [8].

Death is caused by heart failure as well as complications such as hypertensive pulmonary vascular disease and infective endocarditis, so early surgical intervention is advocated for these patients [9]. Prognosis is poor without treatment. Approximately 65% of untreated patient die before 6 months, and up to 90% die before one year of age [10]. Early surgical repair (before 6 months of age) significantly improves outcome [11].

This study aimed to describe the socio-demographic characteristics and the pattern of truncus arteriosus seen in patients with truncus arteriosus at the Obafemi Awolowo University Teaching Hospitals Complex (OAUTHC), Ile-Ife, Nigeria.

2. Methods

A retrospective review of all hospital and outpatient department records of 20 consecutive patients diagnosed with truncus arteriosus at the Paediatric Cardiology Unit (OAUTHC) from January 2017 to January 2021 was undertaken. Pulse oximetry, chest X-ray, electrocardiogram and echocardiography were among the records reviewed. Categorical variables were expressed as percentages/ proportions while continuous variables were expressed as mean ± standard deviation/median. Data was analyzed using the EXCEL statistical package.

Approval for the study was obtained from the Ethics and Review Committee of the Hospital with patient consent waived.

3. Results

There were twenty consecutive patients diagnosed with truncus arteriosus during the period under review. They were equally distributed with 10 males and 10 females, with a male to female ratio of 1: 1. The youngest patient presented at age of 11 days and the oldest at 90 months. The median age at diagnosis was 6.5 months. Oxygen saturation in the children ranged from 36% to 89% with a mean of 72.5± 17.3%.

Table 1. *Characteristics of truncus arteriosus in patients studied.*

Variable	Number (n)	Percentage (%)
Type of truncus arteriosus		
Type I truncus arteriosus	16	80.0
Type II truncus arteriosus	4	20.0
Number of cusps in truncal valve		
Bicuspid truncal valve	1	5.0
Tricuspid truncal valve	12	60.0
Quadricuspid truncal valve	7	35.0
Degree of regurgitation of truncal valve		
No regurgitation	10	50.0
Mild truncal regurgitation	6	30.0
Moderate truncal regurgitation	3	15.0
Severe truncal regurgitation	1	5.0
Origin of coronary arteries		
Normal	17	85.0
Abnormal	3	15.0
Sidedness of the truncal arch		
Left-sided arch	16	80.0
Right-sided arch	4	20.0
Associated abnormalities		
Present	5	25.0
Absent	15	75.0

The weight ranged between 2.65 kg and 17.0 kg with a mean of 7.3± 4.4 kg (median 5.4 kg). There were 16 children (80%) with type I truncus arteriosus while the rest 4 (20%) had type II truncus arteriosus based on the classification by Collett and Edwards.

The truncal valve was tricuspid in 12 (60.0%), quadricuspid in 7 patients (35.0%), and bicuspid in only one (5.0%) patient. Of the 10 (50.0%) patients with regurgitant truncal valves; 6 (60.0%) were mildly regurgitant, 3 (30.0%) were moderately regurgitant while only 1 (10.0%) was severely regurgitant. Four (66.7%) of the mildly regurgitant truncal valves were tricuspid, while 2 (33.3%) were quadricuspid. Two (2) moderately regurgitant truncal valves were quadricuspid while the remaining one (1) was bicuspid.

The only severely regurgitant valve was quadricuspid. However, there were 2 patients with quadricuspid valves that were not regurgitant.

The truncal annulus was dilated in 90% of our patients with z-scores ranging between 3.1-8.58z.

The truncal artery was biventricular in its origin in 16 (80.0%) of the patients while it was univentricular, arising entirely from the left ventricle in 1 (5.0%) child and entirely from the right ventricle in 3 (15.0%) children.

The arch of the aorta was left-sided in 16 (80.0%) of the patients and right-sided in 4 (20.0%) patients. Most of the patients [17 (85.0%)] had a normal origin of the coronary arteries arising from the truncal artery while 3 (15.0) patients had abnormal coronary arteries. Mild pleural effusion was noted in 5 (25.0%) of our patients while the rest did not have any effusion. Six (30.0%) of the children were in heart failure at the time of diagnosis.

Associated anomalies were found in 5 (25.0%) children; 3 (15.0%) of whom had a patent ductus arteriosus, 4 (20.0%) had atrial septal defects; three (3) ostium secundum and one (1) sinus venous type, three (15.0%) had mitral atresia and one (5.0%) tricuspid atresia.

4. Discussion

In this study, 90% of our patients presented outside the neonatal age group and none was diagnosed prenatally. The mean age at diagnosis was 16.8 ± 21.8 months while the median age was 6.5 months. *Animasaun et al.* [12] in Lagos, Nigeria, reported a mean age at diagnosis of 18.4 ± 37.7 months. Under ideal circumstances, most cases of congenital heart defects (CHD) are identified during new-born screening or even in utero by foetal echocardiography [13].

However, a significant percentage of babies with CHD are missed in the early new-born period and are diagnosed after discharge from the hospital during childhood or even during adulthood [13]. Late presentation is one of the major contributors to morbidity and mortality from congenital heart disease in the developing world. Iyer et al. [14] defined late presentation of congenital heart disease as 'presentation late in the natural history of the specific cardiac defect, with consequent transient or irreversible haemodynamic and pathologic alterations that would impact the medical/ surgical approach, risk, and outcome.' Truncus arteriosus is a complex cyanotic congenital heart disease and delay in making the correct diagnosis and instituting treatments early in life affects the overall outcome in children with the disease. Several factors are responsible for late presentation especially in developing nations and these include amongst others: ignorance, late diagnosis and referral, inappropriate medical advice, poor health care system, poor infrastructure, lack of adequate personnel and financial constraints [13].

In Nigeria, health care financing is still mainly out-of-pocket for a significant proportion of the population. The coverage of the health insurance scheme is still very poor and the scheme makes no provision for the care and management of congenital heart diseases.

In this study, males and females were equally affected by the disease, the male to female ratio was 1: 1. *Chen et al.* [3] in a review of the outcome of 54 consecutive patients who had surgical repair of TA reported a male to female ratio of 1.8:1. In a large population of Bohemian children born with congenital heart diseases, girls were more significantly affected with TA than boys (male to female ratio was 1: 1.22) [15]. *Anderson et al.* found more males to be significantly affected by TA than females. *Animasahun et al.* [12] from Nigeria found boys to be more affected, though not significantly than girls (male to female ratio was 1.1: 1). It appears that there is no striking sex predilection with truncus arteriosus in literature.

Varying degrees of cyanosis was present in all the children with oxygen saturation ranging from as low as 34% to 89%.

Eighty percent (80%) of the children we studied had Type I truncus arteriosus according to the classification by Collett and Edward [6]. The rest had Type II TA. *Chen et al.* [3] found Type I truncus arteriosus in two-thirds of the patients they studied. *Tlaskal et al.* [16] reported that over 50% of their patients had Type I truncus arteriosus while *Brown et al.* found Type II TA to be most common in the cohort they studied [4]. Generally, it appears well established in the literature that Type I truncus arteriosus is the most common variant followed by type II, then III and IV [5].

Truncus arteriosus is characterized by a single multi-cuspid semilunar valve with a single vessel, the number of cusps in the truncal valves may vary from 2 to as many as 6 and the valve may be regurgitant or stenotic or both [11, 17, 18].

In two-thirds of the patients in this study, the truncal valve was tricuspid while about a third of the patients had quadricuspid truncal valve. There was only one subject with a bicuspid valve. *Cruz et al.* reported similar findings [19]. *Chen et al.* [3] observed that the truncal valve had three leaflets in more than 80% of the patients they studied, 8% had four leaflets, and a bicuspid valve was observed in 10% of the patients. Other authors have reported that the truncal valve is often tricuspid [4, 5, 17].

Dilatation of the truncal valve was a near-constant feature in all our patients. Often the truncal root is dilated with poorly developed sinuses [5]. Regurgitation of the valve which ranged mostly from mild to moderate was noted in half of the patients we studied, only one patient had severe regurgitation. Other studies [3, 4] have also reported similar findings. Truncal valve incompetence may be due to thickened and dysplastic cusps, prolapse of cusps or fusion of cusps, unequal cusp size, and dilatation of the annulus [5]. Stenosis of the truncal valve is often present when the cusps are nodular and dysplastic. It has been advocated that the truncal valve be replaced whenever abnormalities of the valve are present [5].

In this study, the single trunk was biventricular in origin in 80% of the patients studied. The remaining 20% were univentricular in origin (three from the right ventricle and only one from the left). It has been reported that the truncus arteriosus is of biventricular origin in 68% to 83% of patients.

It arises from the right ventricle in 11% to 29% of patients, and in 4% to 6% from the left ventricle [5].

The truncal arch was right sided in a fifth (20%) of the patients studied. This was similar to the observation by *Tlaskal et al.* [14] who reported the presence of a right aortic arch in 23% of the subjects studied. *Calbalka et al*. [4] noted that a right aortic arch in truncus arteriosus occurs in 21% to 36% of patients.

Associated anomalies reported among the patients we studied included patent ductus arteriosus (15.0%) atrial septal defects (ostium secundum [15%], and sinus venous type [5%], major aortopulmonary collateral arteries (10%), left superior vena cava (5%), mitral atresia (15%), tricuspid atresia (5%), common atrium (5%).

Tlaskal et al. [14] noted the following as anomalies in children with TA: left superior vena cava (7%), truncal valve stenosis (5%), right pulmonary artery stenosis (5%), aberrant right subclavian artery (5%), agenesis of the left pulmonary artery with left lung hypoplasia, mitral stenosis, tricuspid stenosis, partial anomalous pulmonary venous connection, unroofed coronary sinus and hypoplastic aortic arch (1%).

According to *Cabalka et al.* [4], common anomalies seen in children with truncus arteriosus are right aortic arch, interrupted aortic arch, patent ductus arteriosus, unilateral absence of a pulmonary artery, coronary ostial anomalies, and an incompetent truncal valve.

A total or partial anomalous pulmonary venous connection has also been described in persons with TA. Rare associated anomalies that have been reported include tricuspid atresia and mitral atresia [4].

Biventricular hypertrophy, dilation of ventricular chambers and pulmonary arterial hypertension were common secondary complications among our patients.

It is thought that biventricular hypertrophy and dilation of ventricular chambers occur in the presence of truncal valve insufficiency [4]. The prolonged exposure of the pulmonary vascular bed to systemic arterial pressure results in pulmonary arterial hypertension.

5. Conclusion

Truncus Arteriosus is a critical cyanotic congenital heart disease. Delayed diagnosis and limited centers for surgical correction remains a problem in our environment. Type I is the most common variant in our environment and associated anomalies include patent ductus arteriosus, atrial septal defect and mitral atresia.

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