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Tricuspid Atresia: A Rare Cause of Cyanosis among Nigerian Children

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Authors' contributions

This work was carried out in collaboration between all authors. Author BAA designed the study, wrote the protocol and wrote the first draft of the manuscript. Authors ADMW and FBT managed the literature searches, performed the analysis. Author HOG managed the experimental process. Author BAA supervised the overall write up. All authors read and approved the final manuscript.

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ABSTRACT

Background and Objective: There is a dearth of literature on tricuspid atresia among African people. The current study aims to document the clinical profile and prevalence of tricuspid atresia amongst children in a tertiary hospital in South Western Nigeria.
Methods: A prospective review of all consecutive cases of tricuspid atresia diagnosed by echocardiography at the Lagos State University Teaching Hospital (LASUTH) between January 2007 and December 2015. The hospital is located in South Western Nigeria. The subjects were all children with an echocardiographic diagnosis of tricuspid atresia made at the study center. Data were analyzed using Statistical Package for Social Sciences (SPSS) version 20. Level of significance set at $p< 0.05$.

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Results: The prevalence of tricuspid atresia in the study population was 4.6 per 100,000 population of children who presented at the study center during the study period. The prevalence of tricuspid atresia amongst the children with congenital heart disease was 1.3% while its prevalence among children with cyanotic congenital heart disease was 4.3%. Male to female ratio of 1.5:1. The most common class of tricuspid atresia found in this study was class 1C. All but one presented with cyanosis. Surgical intervention was not available in the local environment.

Conclusion: Tricuspid atresia is a rare cause of cyanosis among Nigerian Children; the prevalence of tricuspid atresia in the present hospital based study was lower than the population prevalence in developed countries. There was a male predominance and cyanosis was the most common mode of presentation. The most common class and subtype was 1C. In the sub-region, non-operated cases up to 12 years of age are also reported.

Keywords: Cyanosis; children; Africa; Nigeria; Tricuspid; Atresia.

1. INTRODUCTION

Tricuspid atresia (TA) is recognized worldwide as the third most common cyanotic congenital heart disease after Tetralogy of Fallot and Transposition of the great arteries [1,2] affecting approximately 1 in 10,000 live births [3,4]. Tricuspid atresia results from abnormal development of the tricuspid valve in-utero, which results in non-communication between the right atrium and right ventricle. This defect may be muscular or fibrous depending on the timing of the aberration in-utero [2].

Tricuspid atresia is characterised by failure of de-oxygenated blood from the right atrium to have direct access to the right ventricle and pulmonary arteries for oxygenation in the lungs. For extra-uterine survival, there is usually a communication between both atria for blood to shunt to the left side of the heart. There is also communication between the left ventricle and the pulmonary arteries, and this may be through a ventricular septal defect and or a patent ductus arteriosus. Several classifications of tricuspid atresia have been put forward over the years, presently the acceptable classification based on the relationship between the ventricles and the great vessels is often used [1].

As a result of the defects in patients with tricuspid atresia, the infant becomes cyanosed soon after birth. Other presenting features depend on the associated cardiac defects [1]. Mortality is inevitable without timely surgical intervention. Up to 80 – 90% of the patients who do not have any surgical intervention die at the end of the first year [5,6].

Tricuspid atresia cuts across all geographical locations, race, and gender. The incidence is unknown, but the prevalence amongst another congenital heart disease is 2.9% and 1.4% in autopsy and clinical series respectively in the United States [3]. Internationally, the prevalence is between 1.3 and 2.3% [2]. In Africa, there is a dearth of literature on tricuspid atresia, a report from South Africa documented a prevalence of 1.2-1.4% [7]. Hospital-based studies on the prevalence of congenital heart diseases in children in Nigeria have reported the prevalence of tricuspid atresia to be 1.2 - 4.6% [8-11]. All the reports on tricuspid atresia from Nigeria and sub-Saharan Africa have been part of larger studies on congenital heart disease in children.

To the best of the authors' knowledge, there are no reports from Nigeria, which has been specific on tricuspid atresia in children. The aim of the current study was to document the clinical profile and prevalence of tricuspid atresia amongst children in a tertiary hospital in South Western Nigeria. The findings of this study will add to the body of knowledge on tricuspid atresia both nationally and internationally.

2. METHODS

This study is part of a large study carried out by the Department of Paediatrics, Lagos State University Teaching Hospital, between January 2007 and December 2015. The hospital is located in South Western Nigeria. The Department of Paediatrics is an 84 bedded unit with a Paediatric cardiologist in charge of the cardiology unit. The cardiology unit has facility for ECG, Echocardiography, and Cardiac catheterization. Children were referred from within the state and the sub-region for cardiac evaluation. All children with an

echocardiographic diagnosis of tricuspid atresia were recruited. Echocardiography was performed using a 2-D echocardiography machine with facility for coloured Doppler and M-mode. It is a GE Vivid Q echocardiography machine with reference number 14502 WP SN 2084. Variables of interest including age, gender, clinical presentation, associated intra-cardiac defects and percentage of tricuspid atresia amongst other congenital heart diseases and in the study population were all documented. The analysis was done using Statistical Package for Social Sciences version 20.1. Descriptive statistics was used to determine the mean, standard deviation and median. Variables that were normally distributed were summarized with mean, while skewed data were summarized with median. The relationship between variables was tested with Student t test and Chi square for quantitative and qualitative variables respectively. The level of significance set at $p< 0.05$.

2.1 Ethical Disclosures

The authors declare that no patient data appeared in this write-up. No experiments were performed on the patients for this article.

3. RESULTS

3.1 Prevalence of Tricuspid Atresia

A total of 326,662 children were seen at the Paediatrics department during the study period. Out of those, 15 patients had echo diagnosis of Tricuspid Atresia. The prevalence of tricuspid atresia in the study population was 4.6 per 100,000 population of children who presented at the study centre during the study period. A total of 1,123 children had congenital heart disease. Hence, the prevalence of tricuspid atresia amongst the children with congenital heart disease was 1.3% while its prevalence among children with cyanotic congenital heart disease was 4.3%. Fig. 1 shows the electrocardiographic finding in a patient with tricuspid atresia while Fig. 2 shows 2- dimensional echocardiographic feature in tricuspid atresia.

3.2 Demographic Characteristics of Subjects

There were nine males and six females with a male to female ratio of 1.5:1. The children were aged, 1 month to 12 years with a mean age and standard deviation of 38.6±52.75 months and median of 7 months. The age range for the males and females were 2 months to 12 years

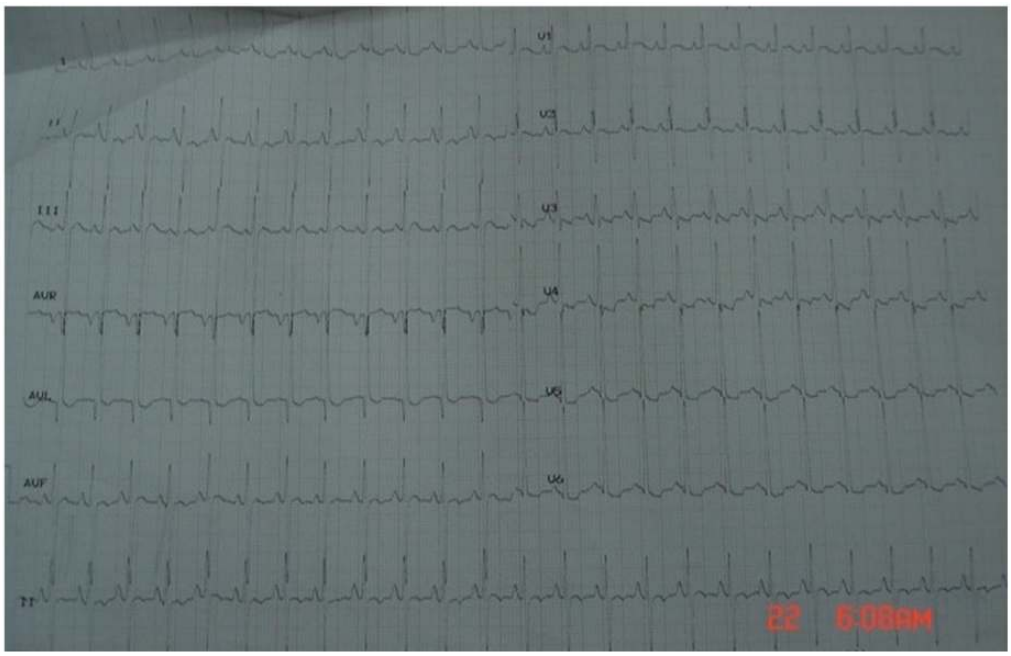

Fig. 1. Electrocardiography of an infant with tricuspid atresia showing left axis deviation, tall p-waves in lead II, left ventricular hypertrophy

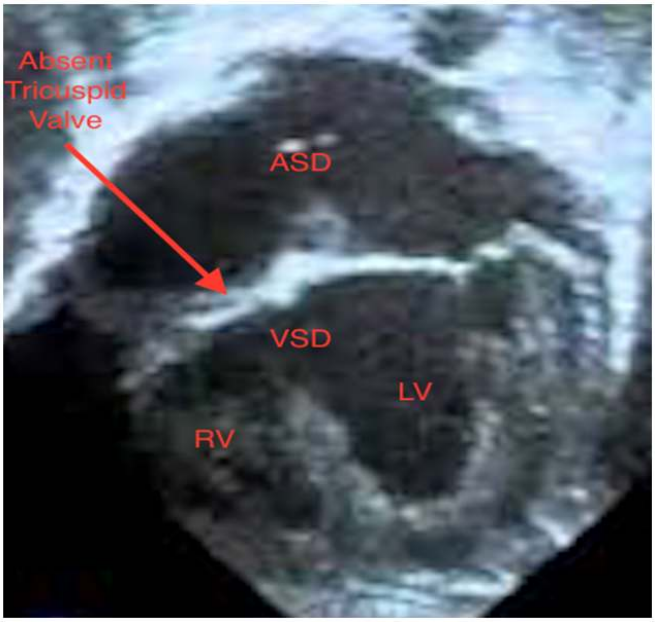

Fig. 2. 2-dimensional echocardiographic image of a child with tricuspid atresia
RV: Right ventricle; LV: Left ventricle; ASD: Atrial Septal Defect; VSD: Ventricular Septal Defect

Table 1. Demographic characteristics of the patients

Variable	Total (%)	Males. n (%)	Females. n (%)	P
Gender	15	9 (60)	6 (30)	0.271
Age group				
≤ 6 months	6 (40)	4	2	0.363
6 months to ≤1 yr	2 (13.3)	1	1	0.363
>1 yr to ≤5 yrs	4 (26.7)	1	3	0.285
>5 yrs to ≤10 yrs	0 (0)	0	0	
>10 yrs	3 (20)	3	0	0.067
Mean age (months)	38.6±52.75	44.61 (64.97)	22.08 (22.07)	
Median age (months)	8	7	14.50	0.864

and one month to 50 months (4.2 years) respectively. The mean and median age for the males and females were 44.61±64.97 months and seven months, 22.08±22.07 months 14.5 months respectively. There was no significant difference in the age at diagnosis across the gender, (p= 0.864). The ages were divided into subgroups. The modal age at diagnosis was ≤6 months. There was no significant difference in the distribution of the age between the genders.

3.3 Associated Cardiac Defects and Types of TA

All patients had absent tricuspid valves with inter-atrial connections. The inter-atrial connections were Patent Foramen Ovale (PFO), Sinus venosus and Secumdum ASDs. The right atrium was dilated in all the patients. The right ventricle was hypoplastic in eight (8) patients and hypertrophic in one patient. The patient that had the hypertrophic right ventricle had truncus arteriosus. The type and classification of tricuspid atresia based on the great vessel relationship and characteristics of the pulmonary artery are highlighted in Table 2. The most common class of tricuspid atresia found in this study was class 1, (ventriculoarterial concordance), occurring in 2/3rd (66.7%) of the patients. Of the 10 patients with class 1, class 1C (normal pulmonary artery) was the most common subtype. Class 2, (D-TGA) was documented in 4 patients (26.7%) while only one patient (6.7%) had class 4, (truncus arteriosus). There was no class 3 in the study population.

3.4 Clinical Presentation and Outcome

All but one patients was cyanosed at presentation. The only patient that was not clinically cyanosed was desaturating in room air and had an oxygen saturation of 86%. That was a patient with truncus arteriosus. One patient presented at one month of age with cyanosis and heart failure, with a chest radiograph revealing dextrocardia. That patient had class 1A tricuspid atresia. Most of the patients either died or were lost to follow-up. One of the patients was referred for surgery at the age of six years but was inoperable because of obstructive pulmonary vascular disease. He was followed up at the cardiology clinic regularly but died about two years later from features suggestive of intracranial haemorrhage. Unfortunately, autopsy was not done to confirm this because the parents did not give consent.

3.5 Clinical Management

All the patients were commenced on oral medications as needed based on their clinical presentations and co-morbidities. Medications ranged from diuretics, angiotensin converting enzyme inhibitors to beta blockers for patients with congestive cardiac failure and right ventricular outflow obstruction respectively. Common co-morbidities were growth retardation and respiratory tract infection. These were managed appropriately with other treatment modalities and clinical specialists as required.

All the patients required surgical corrections. This was not possible in Nigeria, thus they were referred for treatment abroad. 90% of the patients could not afford care abroad, thus they succumbed before funds could be sourced for surgical corrections.

4. DISCUSSION

The prevalence of tricuspid atresia in the present study is lower than the population prevalence in developed countries [3,4]. While the denominator of the population prevalence from previous reports were live births, the present study had children ≤13 years. Possible explanation for the lower prevalence in the present study may be because this is a hospital-based study involving children up to 13 years who only presented to the hospital on account of an ill health. Thus children who didn't present to the hospital were missed out. Secondly, some patients with tricuspid atresia may have been missed in the first month of life before an echocardiography was done. Thirdly, some patients who may have died from suspected cyanotic heart lesions did not have post-mortem done. The most common reasons for refusal to have post mortem done on deceased children are cultural and religious.

Table 2. Characteristic of and classification of TA

S/N	Relationship of great vessels and gender of the patients		Pulmonary artery	Right ventricle	IVS	Class
1	Concordant	M	Normal	Hypoplastic	PM	1C
2	D-TGA with DORV	M	Atretic	-	SA	2A
3	Concordant	M	Normal	-	-	1C
4	Concordant	F	Normal	Hypoplastic	PM	1C
5	D-TGA	M	Normal	Hypoplastic	-	2C
6	Concordant	M	Stenosed	Hypoplastic	-	1B
7	Concordant	F	Stenosed	-	-	1B
8	Concordant	M	Normal	Hypoplastic	-	1C
9	D-TGA	M	Stenosed	-	Inlet	2B
10	Truncus Arteriosus	F	Stenosed	RVH	-	4B
11	D-TGA	F	Normal	Hypoplastic	SA	2C
12	Concordant	F	Normal	Hypoplastic	SA	1C
13	Concordant	M	Atretic	Hypoplastic	-	1A
14	Concordant with DORV	F	Atretic	-	-	1A
15	Concordant	M	Normal	-	Muscular	1C

D-TGA = Dextro-Translocation of the great arteries, DORV = Double Outlet Right Ventricle, IVS = Intraventricular septum, PM = Perimembranous, SA = Sub-aortic, RVH = Right ventricular hypertrophy.
M = Male, F = Female

Thus the prevalence of tricuspid atresia given herein may be an underestimate of the true prevalence in the sub-region given that the present study is hospital based. A careful literature search revealed that there is no data on population prevalence of tricuspid atresia in the sub-region.

The international prevalence of tricuspid atresia amongst congenital heart diseases varies between 1.2-1.4% [3]. The prevalence of tricuspid atresia amongst the congenital heart diseases herein documented is similar with reports from the literature. Some authors have documented rates as high as 4.6% in Nigeria. The reports in Nigeria have a duration of study that ranged from 2 to 10 years. Antia [8] in 1973 reported a prevalence of 1.2% over a 10year period which is more closely related to results in the present study. The difference between the present study and the earlier research by Antia [8] is that while the present study consists only of clinical series, the earlier study considered live patients and autopsy reports. Concerning the prevalence of tricuspid atresia amongst the cyanotic congenital heart diseases, in the present study, tricuspid atresia was documented in less than 5% of the cyanotic congenital heart diseases. The prevalence in that regard is low compared to reports from the USA where it is known as the third most common cyanotic congenital heart disease [12]. There are no reports in that regard within the sub-region. Thus it was difficult to compare the prevalence of tricuspid atresia amongst the cyanotic congenital heart diseases in sub-Saharan Africa.

Given that tricuspid atresia is a cyanotic congenital heart disease, the patients present with cyanosis soon after birth. The degree of pulmonary blood flow also affects the timing of presentation [1]. Patients with pulmonary oligemia present earlier with cyanosis while those with pulmonary plethora present later with congestive cardiac failure and cyanosis [1]. Not surprising, almost all the patients in the present study were cyanosed at presentation. The youngest age at presentation was one month of age and the oldest patient was 12 years old. It is known that up to 80% of the patients are symptomatic and present at the end of the first month of life [6,13] and only 10-20% of unoperated patients survive till the end of the first year [2]. It was thus surprising to report un-operated cases after infancy and at 12 years of age. A possible explanation may be because up to half of the patients in this study had normal pulmonary arteries. Tricuspid atresia is slightly more common in males [3]. The male preponderance is even more, up to twice the number of females, in the subset of patients with transposition [6]. In the present study the males were more than the females, and out of the four patients with transposition of the great arteries, three were males. The findings thus corroborate earlier reports.

Tricuspid atresia can be classified based on the relationship of the great arteries and the characteristic of the pulmonary artery into four main classes and three subtypes [13]. In the present study, the most common class was 1C. Internationally, the most common type is class 1 [14] which was noted in the present study. The most common subtype is class 1B, but this was different from the report in the present study where 1C was the most common. All the patients had a muscular defect across the right atrioventricular valve. The finding in this regard is in not surprising given that the muscular subtype is the most common type, and it has been documented in up to 89% of cases [15].

The diagnosis of tricuspid atresia can be made in-utero with fetal echocardiography and electrocardiogram. In the extra-uterine life, a combination of clinical features with electrocardiography and echocardiography can clinch the diagnosis. In developing countries, intra-uterine echocardiography is hardly ever done, and cases are thus missed. In the extra-uterine life, the use of echocardiography is becoming increasingly common, and thus, cases are identified. But the cases identified may be a tip of the iceberg given that access to health care is not yet optimal. For the patients that are diagnosed, the standard of care which comprise surgical intervention is not readily accessible and available. Thus, patients are diagnosed but die before optimal care is given.

Limitations of the present study includes the fact that it was hospital based. This may have resulted in a lower prevalence in the sub-region compared with documented values in other regions. Ideally a community based study may reflect a more accurate prevelence than the present study. A coummity study will be more expensive to carry out and the present study aimed to document hospital prevelence of tricuspid atresia. Another limitation was that, the prevalence of tricuspid atresia could not be

documented in all newborns. This is because in the present study site, there are no inborn deliveries. Thus the prevalence of tricuspid atresia amonsgt all newborn could not be documented in the present study.

Despite the above limitations, this study has presneted the hospital prevalence and clinical profile of children with Tricuspid atresia. This information is relevant because there are no reports of tricuspid atresia in the sub-region. Given the information that we have highlighted herein, it may be necesaary in the future, to carry out a multicentred and or commuinity study on Tricuspid Atresia and findings may be compared with this report.

5. CONCLUSION

In conclusion, we documented a prevalence of 4.6 per 100,000 children for tricuspid atresia among the subjects, 1.2% amongst those with congenital heart disease and 4.2% amongst those with cyanotic congenital heart diseases. There were a male predominance and the most common class, and subtype was 1C. In the sub-region non-operated cases up to 12 years of age are also reported.

CONSENT

It is not applicable.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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