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Original article

# Complex congenital heart defects in children in a resource constraint setting

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## Abstract

**Background:** Complex congenital heart defects (CHD) are among the non-infectious causes of childhood morbidity and mortality in developing countries, accounting for 25-30% of intensive care usage in some centers. In Abuja, the capital of Nigeria, there has been an upsurge in cases of complex CHD, thus creating the need for this study.

**Objectives:** The study aimed at describing the incidence and outcome of complex CHD in children seen at the University of Abuja Teaching Hospital, Gwagwalada, Abuja.

**Methods:** A prospective study of patients referred to the Pediatric Cardiology Unit of the hospital between 2013 and 2018 was performed.

**Results:** Of the 1,420 patients examined, 344 (24.2%) had CHD; 55 (16.0%) of these belong to the complex category with a male:female ratio of 1.3:1. The incidence of complex CHD is 1.1 per 1,000 live births. Tetralogy of Fallot (TOF) was the commonest form of complex heart diseases. Patients with atresic valves tend to present in the neonatal and the immediate postnatal period, while those with TOF present much later. Only 8 (14.5%) had surgical interventions, 21 (38.2%) could not afford surgery, 14 (25.5%) were lost to follow up and 10 (18.2%) died. Patients that had need of emergency cardiac surgery had the worst prognosis. Two patients with truncus arteriosus presented too late and were classified inoperable. Financial constraint and lack of facility for cardiac surgeries contributed negatively to the outcome of patients with complex heart defects.

**Conclusions:** Complex CHD occurred in this community with similar incidence rates as in the developed countries. The predominant complex heart defect in the locality is TOF. Defects with atresic valves carry the worst prognosis in this environment due to lack of the wherewithal to manage the condition.

## Keywords

Children, complex heart defects, outcome, resource constraint, Nigeria.

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## Introduction

Congenital heart defects (CHD) are among the non-infectious causes of morbidity and mortality in children in developing countries with an estimated incidence of 8-12 per 1,000 live births [1, 2]. Generally, CHD has been classified based on pathophysiological processes (into simple and complex) and anatomical defects (into shunt defects, obstructive lesions, conotruncal abnormalities and complex disorders) [3]. When a congenital anomaly involves parts of the heart that are critical for the sustenance of a patient's life, it is considered to be complex [4]. Unlike the simple CHD, most complex heart diseases present as emergencies requiring urgent and prompt intervention. Though considered uncommon, complex CHD has been recorded in 0.64-2.3 per 1,000 live births in a recent publication and account for up to 25% of intensive care facility usage in some centers [5, 6]. Improvement in diagnosis and management have significantly improved the life expectancy in babies born with the disease [7, 8].

A dedicated healthcare facility is often required to handle the challenges involved in ensuring survival of majority of children with very complex heart lesion. The establishment of such facilities will however be dependent on the knowledge of the pattern of CHD in a given population. Therefore, this study was focused on the incidence of complex CHD and outcome of the disease in Abuja, and highlights the challenges with the management of CHD in an economically disadvantaged society.

## Methods

A prospective study of patients with complex CHD in the Paediatric Cardiology Unit of the University of Abuja Teaching Hospital, over a 5-year period spanning August 2013 to July 2018, was done. Information obtained included type of complex heart lesion, age at diagnosis, the gender of the patient and the outcome of management. A heart lesion was considered complex if it posed a threat to the survival of the child and included any of the following: tetralogy of Fallot (TOF), transposition of great arteries (TGA), atresia of valves, single ventricle physiology (SV), total anomalous pulmonary venous drainage (TAPVD), truncus arteriosus (Trunc A), double outlet right ventricle (DORV) or double inlet left ventricle (DILV) and dextrocardia with intracardiac abnormalities.

Data was analysed using frequency tables and Chi-square statistic test. A p-value $< 0.05$ was set as point of statistical significance.

## Results

A total of 1,420 children were referred to the unit over the study period, 344 (24.2%) had congenital heart disease, of which 55 (16.0%) were of the complex type. Thirty-one (56.4%) of the 55 complex CHD were found in males while 24 were in females giving a male:female ratio of 1.3:1

TOF was diagnosed in 23 (41.8%) patients with complex cardiac anomaly while TGA, Trunc A, pulmonary atresia (PA) and SV occurred in 9.1%, respectively. Details of the distribution of the complex heart lesion are displayed in **Tab. 1**.

**Table 1.** Frequency of complex congenital heart defects (CHD) in children.

| Types of heart defects | Number of cases | Percentage |
|---|---|---|
| **TOF** | 23 | 41.8 |
| **TGA** | 5 | 9.1 |
| **Trunc A** | 5 | 9.1 |
| **PA** | 5 | 9.1 |
| **DORV** [a] | 5 | 9.1 |
| **SV** [b] | 5 | 9.1 |
| **TA** | 3 | 5.5 |
| **Common atrium** | 2 | 3.6 |
| **Dextrocardia with other anomalies** | 2 | 3.6 |

[a]One of the patients with DORV had Taussig-Bing anomaly; [b]SV included hypoplastic left heart syndrome (HLHS) (2), hypoplastic right heart syndrome (HRHS) (1), Holmes heart (1), congenital mitral atresia (MA) (1).
TOF: tetralogy of Fallot; TGA: transposition of great arteries; Trunc A: truncus arteriosus; PA: pulmonary atresia; DORV: double outlet right ventricle; SV: single ventricle physiology; TA: tricuspid atresia.

*Incidence of complex heart defects*

There were 10,065 live births in the hospital during the study period. Eleven (0.11%) of them had complex heart defects, thus the hospital incidence of complex CHD is 1.1 per 1,000 live births. The incidence of TOF was 0.3 per 1,000 live births while that of TGA, Trunc A and PA were 0.2 per 1,000 live births each. The rarer complex CHD such as hypoplastic left heart syndrome (HLHS) and mitral atresia (MA) occurred in 0.1 per 1,000 live births (**Tab. 2**).

*Age at presentation/diagnosis*

Diagnosis was made in the neonatal period in 3 patients with TOF, 2 patients with TGA and 1 patient each for the more complex heart lesions. On the overall, 15 (27.3%) of the complex heart lesions were diagnosed in the neonatal period, while 16 (29.1%) were diagnosed between 1 and 6 months of life. As shown in **Tab. 3**, only 4 (7.3%) of the patients presented later than 5 years. Thirteen of the 23 patients with TOF presented after infancy while 4 of the 5 patients with Trunc A were diagnosed between 1 and 6 months of life (**Tab. 3**). With further analysis, the non-TOF complex heart defects tended to be diagnosed earlier than 6 months of life compared to the TOF cases ($\chi^2 = 15.67$, $p = 0.00008$).

*Outcome of complex congenital heart defects*

Of the 55 patients with complex heart defects, 8 (14.5%) have had surgical interventions and are doing well, 21 (38.2%) could not afford the cost of surgery and have remained regular on palliative medical therapy with either chronic antifailure drugs or recurrent partial saline exchange transfusion. Fourteen (25.5%) patients were lost to follow up while 10 (18.2%) deaths were recorded. In 2 patients, the heart defects were inoperable due to late presentation. Details of outcome of patients with complex CHD are shown in **Tab. 4**.

**Table 2.** Incidence of complex congenital heart defects (CHD) among babies born in the hospital.

| Types of defects | No. of cases among inborn, n (%) | No. of cases referred from other hospitals, n (%) | Total | Incidence per 1,000 LB [b] | Overall incidence [b] |
|---|---|---|---|---|---|
| **TOF** | 3 (13.0) | 20 (87.0) | 23 | 0.3 | 1 in 3,300 LB |
| **TGA** | 2 (40.0) | 3 (60.0) | 5 | 0.2 | 1 in 5,000 LB |
| **Trunc A** | 2 (40.0) | 3 (60.0) | 5 | 0.2 | 1 in 5,000 LB |
| **PA** | 2 (40.0) | 3 (60.0) | 5 | 0.2 | 1 in 5,000 LB |
| **HLHS** | 1 (50.0) | 1 (50.0) | 2 | 0.1 | 1 in 10,000 LB |
| **MA** | 1 (100) | - | 1 | 0.1 | 1 in 10,000 LB |
| **Overall** | 11 (26.8) | 30 (73.2) | 41 [a] | 1.1 | 1.1 in 1,000 LB |

[a] The remaining 14 defects were seen only in patients referred from other hospitals; [b] incidence was calculated for inborn patients (LB in the hospital during the study period = 10,065).
LB: live births; TOF: tetralogy of Fallot; TGA: transposition of great arteries; Trunc A: truncus arteriosus; PA: pulmonary atresia; HLHS: hypoplastic left heart syndrome; MA: mitral atresia.

**Table 3.** Distribution of complex congenital heart defects (CHD) according to the age at diagnosis.

| Age at diagnosis | Types of complex CHD | | | | | | |
|---|---|---|---|---|---|---|---|
| | TOF, n (%) | TGA, n (%) | Trunc A, n (%) | PA, n (%) | TA, n (%) | DORV, n (%) | Others, n (%) |
| **Neonate** | 3 (13.0) | 2 (40.0) | - | 1 (20.0) | 1 (33.3) | 2 (40.0) | 6 (66.7) [a] |
| **1-6 months** | 2 (9.0) | 1 (20.0) | 4 (80.0) | 3 (60.0) | 2 (66.7) | 2 (40.0) | 2 (22.2) [b] |
| **7-12 months** | 5 (22.0) | 1 (20.0) | - | 1 (20.0) | - | 1 (20.0) | - |
| **1-5 years** | 10 (43.2) | 1 (20.0) | 1 (20.0) | - | - | - | - |
| **≥ 6 years** | 3 (13.0) | - | - | - | - | - | 1 (11.1) [c] |
| **Total** | 23 | 5 | 5 | 5 | 3 | 5 | 9 |

[a] Included mitral atresia (MA) (1), hypoplastic left heart syndrome (HLHS) (2), hypoplastic right heart syndrome (HRHS) (1), Taussig-Bing (1) and common atrium (1); [b] included dextrocardia with severe pulmonary stenosis (1) and Holmes heart (1); [c] dextrocardia with situs inversus.
CHD: congenital heart defects; TOF: tetralogy of Fallot; TGA: transposition of great arteries; Trunc A: truncus arteriosus; PA: pulmonary atresia; TA: tricuspid atresia; DORV: double outlet right ventricle.

**Table 4.** Outcome of management of patients with complex congenital heart defects (CHD).

| Heart defects (n) | Associated conditions | No. of cases | Treatment given | Outcome/remarks |
|---|---|---|---|---|
| TOF (23) | Polycythaemia | 12 | Oral propranolol Partial saline transfusion | Occasional hypercyanotic spells. Financial constraints deferring surgical intervention |
| | Severe financial constraints | 7 | Oral propranolol | Lost to follow up |
| | Nil | 3 | Surgical correction in India | Stable with residual pulmonary regurgitation in two patients |
| | Nil | 1 | Modified Blalock Taussig in Ghana | Developed postoperative chylothorax. Lost to follow up |
| TGA (5) | Spongy myocardium | 1 | Had Glenn procedure in India | Arterial switch deferred due to spongy myocardium |
| | Large VSD | 2 | Antifailure therapy | Failure to thrive. Couldn't access healthcare abroad due to lack of fund |
| | ASD, PDA, VSD | 2 | Antifailure therapy | Lost to follow up |
| Trunc A (5) | Diaphragmatic hernia | 1 | Antifailure regimen | Died within 1 week of life |
| | Endocarditis | 1 | Antibiotics, antifailure therapy | Died at 3 months of life |
| | Late presentation | 2 | Antifailure regimen | Inoperable due to late presentation |
| SV (5) | MA and omphalocoele major | 1 | Acute antifailure regimen | Died in the 2nd week of life |
| | HLHS | 1 | Enalapril, oral $PGE_1$ | Died at home at 3 weeks of life |
| | Holmes heart | 1 | Antifailure regimen | Lost to follow up |
| | HRHS | 1 | Antifailure regimen | Stable at last visit |
| PA (5) | Intact interventricular septum | 2 | Needed urgent atrial septostomy but not available | Died at 2 weeks of life |
| | Large VSD, one of whom developed endocarditis | 3 | Antibiotics were given to patient with endocarditis | Patient with endocarditis died, 1 patient had recurrent polycythaemia and intermittent partial saline exchange transfusion, and 1 was lost to follow up |
| DORV (5) | Polycythaemia Iron deficiency Single parenthood | 1 | Recurrent partial saline exchange transfusion | Could not afford the cost of surgery |
| | Nil | 2 | Corrective surgery done in India | Thriving well |
| | Severe financial constraints | 2 | Antifailure regimen | Lost to follow up |
| Dextrocardia (2) | Severe pulmonary stenosis, pneumonia | 1 | Antibiotics | Died suddenly at 8 weeks of life |
| | Situs inversus, bradycardia | 1 | Atropine | Awaiting pacemaker insertion |
| TA (3) | Severe sepsis | 1 | Antibiotics, antifailure therapy | Died at 3 months of life |
| | Nil | 2 | Antifailure therapy | Lost to follow up |
| Common atrium (2) | AVSD, Ellis van Creveld syndrome | 1 | AVSD correction in India with permanent pacemaker | Child is growing, developed ricket |
| | Heart failure | 1 | Antifailure therapy | Child is thriving, awaiting surgery |

TOF: tetralogy of Fallot; TGA: transposition of great arteries; Trunc A: truncus arteriosus; SV: single ventricle physiology; PA: pulmonary atresia; DORV: double outlet right ventricle; TA: tricuspid atresia; VSD: ventricular septal defect; ASD: atrial septal defect; PDA: patent ductus arteriosus; MA: mitral atresia; HLHS: hypoplastic left heart syndrome; HRHS: hypoplastic right heart syndrome; AVSD: atrioventricular septal defect.

## Discussion

The incidence of complex CHD in this study is 1.1 per 1,000 live births, an incidence rate similar to the 0.9-1.1 per 1,000 live births reported in Beijing, China [9] and Anatolia, Turkey [10] but lower than the 1.5 per 1,000 live births in Taiwan [11] and the 1.99 per 1,000 live births in Massachusetts, USA [12]. Though the incidence of complex CHD shows mild regional variations, it is generally similar across the globe, clustering around 1-2 per 1,000 live births.

TOF constituted the highest number of complex CHD seen in this study, accounting for 41.8% of all cases seen and has an incidence of 0.3 per 1,000 live births. The predominance of TOF among complex CHD has been documented in studies from other parts of Nigeria [13, 14], and the world at large [5, 6, 9-11, 15]. The review of the Guangdong Registry of Congenital Heart Disease by Qu et al. [15] as well as the European Registry of Congenital Anomalies (EUROCAT) documented the same incidence of 0.3 per 1,000 live births for TOF as seen in this study.

The other less common CHD such as TGA, PA, tricuspidal atresia (TA) and HLHS occur with the same incidence rates as reported in technologically advanced countries like China and various parts of Europe [15]. Thus further corroborating the general perception on the similarity in incidence of CHD across countries of the world.

The fact that complex CHD are often symptomatic and present early in life makes the diagnosis easier and could have accounted for the relatively uniform incidence rates all over the world [15].

The more complex the heart lesions, the higher the tendency to earlier diagnosis. Thus heart lesions with more complex defects were diagnosed much earlier than TOF in the present study. TOF tends to manifest symptoms at a relatively older age in most cases except for those with very severe right ventricular outflow tract obstruction. On the contrary, patients with atresic valves (PA, TA, MA) clustered around the neonatal and immediate post-neonatal period. This observation highlights the need for facilities that support timely neonatal cardiac surgeries in this community. Otherwise mortality from complex CHD will be alarming.

The outcome of complex CHD in this study leaves much to be desired. Majority of patients with atretic valves died due to absence of facilities for open heart surgery in the locality. So far only 14.5% of the patients with severe heart defects have had surgical intervention done abroad. The remainders are either on palliative medical therapy or lost to follow up, while some have died. Delay in surgical intervention is largely due to financial constraints and, since the National Health Insurance Scheme does not cover cardiac surgeries, parents have the options of paying for the surgeries abroad, or waiting for one of the outreach surgeries by foreign doctors, both of which are not readily available. Thus resulting in most patients being treated palliatively with either recurrent partial saline exchange transfusion or anti-heart failure drugs.

Although there are reports that almost 85% of babies born with CHD attain adulthood in developed countries [6, 7, 16, 17], the situation in our community, as it is in many African countries [14, 15], suggests a contrary view where mortality from complex heart defects is almost unpreventable. Therefore, there is a need for policy makers to establish a centre where babies with cardiac related emergencies could be referred and managed promptly.

## Conclusion

The incidence rates of complex CHD in this community are similar to those reported in more developed countries. TOF is the predominant type of complex heart defects. Defects with atresic valves carry the worst prognosis in this environment due to lack of the wherewithal to manage the condition.

## Declaration of interest

The Authors declare that there is no conflict of interest.

## References

1. Loffredo CA. Epidemiology of cardiovascular malformations: prevalence and risk factors. Am J Med Genet. 2000;97(4):319-25.
2. Hoffman JIE. The global burden of congenital heart diseases. Cardiovasc J Afr. 2013;24:141-5.
3. Bassareo PP, Saba L, Solla P, Barbanti C, Marras AR, Mercuro G. Factors influencing adaptation and performance at physical exercise in complex congenital heart diseases after surgical repair. Biomed Res Int. 2014;2014:862372.
4. Miyague NI, Cardoso SM, Meyer F, Ultramari FT, Araújo FH, Rozkowisk I, Toschi AP. Epidemiological study of congenital heart defects in children and adolescents. Analysis of 4,538 Cases. Arg Bras Cardiol. 2003;80:269-78.
5. Miranović V. The incidence of congenital heart defects in the world regarding the severity of the defect. Vojnosanit Pregl (Engl). 2016;73:159-64.
6. Reller MD, Strickland MJ, Rielble-Colarusso T, Mable WT, Correa A. Prevalence of Congenital heart defects in metropolitan Atlanta, 1998-2005. J Pediatr. 2008;153:807-13.
7. Khairy P, Ionescu-Ittu R, Mackie AS, Abrahamowicz M, Pilote L, Marelli AJ. Changing mortality in congenital heart disease. J Am Coll Cardiol. 2010;56:1149-57.
8. Marelli AJ, Mackie AS, Ionescu-Ittu R, Rahme E, Pilote L. Congenital heart disease in the general population: changing prevalence and age distribution. Circulation. 2007;115:163-72.

9. Yang XY, Li XF, Liix D, Liu YL. Incidene of congenital heart disease in Beijing, China. Chin Med J (Engl). 2009;122: 1128-32.
10. Baspinar O, Karaaslan S. Oran B, Baysal T, Elmali AM, Yorulmaz A. Prevalence and distribution of children with congenital heart diseases in the central Anatolian region, Turkey. Turk J Pediatr. 2006;48:237-43.
11. Wu MH, Chen HC, Wang JK, Huang SK. Prevalence of congenital heart diseases at live birth in Taiwan. J Pediatr. 2010;156(5):782-5.
12. Liberman RF, Getz KD, Lin AE, Higgins CA, Sekhavat S, Markensen GR, Anderka M. Delayed diagnosis of critical congenital heart defects: trends and associated factors. Pediatrics. 2014;13(2): e373.
13. Chinawa JM, Obu HA, Eke CB, Eze JC. Pattern and clinical profile of Children with complex Cardiac anomaly at University of Nigeria Teaching Hospital, Ituku-Ozalla, Enugu State, Nigeria. Niger J Clin Pract. 2013;16:462-7.
14. Sani Mu, Mukhtar-Yola M, Karaye KM. Spectrum of Congenital heart diseases in a tropical environment: an echocardiography study. J Natl Med Assoc. 2007;99:665-9.
15. Qu Y, Liu X, Zhuang J, Cheng G, Mai J, Guo X, Ou Y, Chen J, Gong W, Gao X, Wu Y, Nie Z. Incidence of congenital heart disease: The 9-year experience of the Guangdong Registry of Congenital Heart Disease, China. PLos One. 2016;11(7):e015925.
16. Mocumbi AO, Lamiera E, Yaksh A. Challenges on the management of congenital heart disease in developing countries. Int J Cardiol. 2011;148:285-88.
17. Zühlke L, Mirabel M, Marijon E. Congenital heart disease and rheumatic heart disease in Africa: recent advances and current priorities. Heart. 2013;99:1554-61.