Pattern of congenital heart disease among children presenting to the Uganda Heart Institute, Mulago Hospital: a 7-year review

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Abstract

Background: Congenital heart disease (CHD) is the most common congenital anomaly in children. Over half of the deaths due to CHD occur in the neonatal period. Most children with unrepaired complex heart lesions do not live to celebrate their first birthday. We describe the spectrum of congenital heart disease in Uganda.

Methods: We retrospectively reviewed the data of children with CHD who presented to the Uganda Heart Institute (UHI), Mulago Hospital Complex from 2007 to 2014.

Results: A total of 4621 children were seen at the UHI during the study period. Of these, 3526 (76.3%) had CHD; 1941 (55%) were females. Isolated ventricular septal defect (VSD) was the most common CHD seen in 923 (27.2%) children followed by Patent ductus arteriosus (PDA) 760 (22%) and atrial septal defects (ASD) 332 (9.4%). Tetralogy of Fallot (TOF) and Truncus arteriosus were the most common cyanotic heart defects (7% and 5% respectively). Dysmorphic features were diagnosed in 185 children, of which 61 underwent genetic testing (Down syndrome=24, 22q11.2 deletion syndrome n=10). Children with confirmed 22q11.2 deletion had conotruncal abnormalities.

Conclusion: Isolated VSD and Tetralogy of Fallot are the most common acyanotic and cyanotic congenital heart defects. We report an unusually high occurrence of Truncus arteriosus.

Keywords: Congenital heart disease; children; Uganda.

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Introduction

Congenital heart disease (CHD) is the most prevalent congenital abnormality and a leading cause of childhood mortality1. The estimated prevalence of CHD is 8-12/1000 live births2-4. Without appropriate treatment, about one in three children born with significant congenital heart disease will die within the first month of life5. Unrepaired congenital heart disease is a major cause of heart failure among children in Africa6. We believe that this study is the largest in Africa to report on congenital heart disease amongst children. The aim of this study was to describe the spectrum of congenital heart disease among children who presented to the Uganda Heart Institute.

Methods

We retrospectively reviewed 3526 echocardiography reports of patients with CHD that presented to the Uganda Heart Institute (UHI) between 2007 to 2014. The registry for congenital heart disease was established in 2007 by the pediatric cardiology division as part of the Ministry of Health reporting system and serves as the basis for this study. Makerere University School of Medicine Ethics and Research Committee approved the genetics component of the study.

Study site

The Institute is a 40 bed facility located within the Mulago Hospital complex and has been in existence for 28 years. The UHI has a fully functional operating theatre in addition to a catheterization laboratory. It performs pediatric and adult open-heart surgeries, diagnostic procedures, and offers a variety of specialized services.
and interventional catheterization procedures. Outpa-
tient clinics run on a daily basis. Averagely, 60 to 80 paed-
diatric open heart surgeries are conducted per year. A
paediatric cardiology fellowship program has run since
2010.

Study procedure
Detailed transthoracic echocardiography was per-
formed and interpreted by one of the two pediatric
cardiologists (PL /SL) using standard guidelines7 with
a Sonos 5500 (Philips, Best, Netherlands) and a Philips
IE 33 (Philips, Best Netherlands) for the periods 2007
to 2011 and 2012 to 2014 respectively.
Difficult cases were discussed and a final diagnosis
made by consensus. Digital archiving enabled cases to
be reviewed and discussed with colleagues (CS) from
other centers. Re-evaluation of cases at follow up im-
proved diagnostic accuracy.
Severe congenital Heart disease, CHD was defined as
complex heart abnormalities that were life-threatening.
For example, heterotaxy syndromes, anomalous origin
of the left coronary artery from the pulmonary artery
(ALCAPA) and univentricular heart. Patient demo-
graphics including age, sex, weight and type of congen-
tial heart defect were entered into an Excel spreadsheet
and analyzed using SPSS version 16. Pulse oximetry
readings were available for only a small subset of chil-
dren and not included in the analysis.

Syndromic children
If the child had an obvious syndromic condition based
on clinical examination, they were sent for genetics as-
sessment. Genetics testing was done by a highly experi-
cenced genetics specialist from Washington DC, United
States together with the Ugandan team and the paediat-
ric cardiologist (CS) during some missions. Genetics di-
agnosis was based on clinical presentation of the com-
mon genetics syndromes with associated complications.
These were matched with the echocardiogram diagno-
sis. We secured Institutional Ethical approval from the
Makerere School of Medicine as well as from the Na-
tional Institute of Health (NIH) laboratory in Wash-
gton DC, United States to run detailed microarray and
DNA sequencing for the genetics study but blood sam-

ple were not taken off or sent for analysis. The team
relied on clinical diagnosis by a highly skilled genetics
specialist. Syndromes such as Down's syndrome, 22q11
deletion syndrome, Holt Oram and Williams syndrome
were diagnosed. Children with Holt Oram syndrome
had upper limb abnormalities in addition to the CHD.
A genetics syndrome was diagnosed based on clinical
evaluation, the results were given to the family, and was
counselled by our team, a geneticist and genetic coun-
selor.

Congenital Rubella Syndrome
Congenital Rubella syndrome, CRS was diagnosed
based on clinical features of cataracts, microphthalmia,
microcephaly and hearing impairment. Retrospective
data was obtained from the World Health Organization
-Congenital Rubella (WHO- CRS) Surveillance that was
conducted at the UHI in 2014. CRS was confirmed by
blood samples obtained from the child and mother.
Serum was tested at Uganda Virus Research Institute
(UVRI) for evidence of active rubella virus infection
through identification of rubella-specific IgM antibod-
ies. UVRI is a government parastatal certified by the
American College of Pathologist to conducts research,
surveillance and diagnostics linked to viral etiology and
provides expert advice8.

Results
Overall, 4621 charts were reviewed during the study pe-
riod. A diagnosis of congenital heart disease was made
in 3526 (76.3%) children. The majority 1941 (55%) were
females. Most patients presented during infancy (range
1 day -18 years).
VSD was the most prevalent defect (921) of which
702 children (76%) had perimembranous VSD, and 79
(8.5%) had muscular VSD. PDA was the second most
commonly occurring defect seen in 760 cases (22%).
ASD was present in 332 children (9.4%) with the Os-
tium secundum type occurring in 293 (88%) followed
by the sinus venosus defect, 23 (6.9%).
Tetralogy for Fallot and persistent Truncus arteriosus
were the most common cyanotic heart diseases. Some
defects occurred in small percentages and have been
reported. Children with syndromes in the study were
examined for specific genetic abnormalities.
**Table 1:** A cyanotic heart diseases

| Lesion                          | Number | Overall Percentage in CHD (%) | Mean age (months)/years | Female n (%) |
|---------------------------------|--------|------------------------------|------------------------|--------------|
| Isolated VSD                    | 921    | 26                           | 25(2)                  | 484 (52)     |
| PDA                             | 760    | 22                           | 19(1.6)                | 478 (62)     |
| ASD                             | 332    | 9.4                          | 51(4)                  | 188 (56)     |
| ECD                             | 265    | 8                            | 17(1)                  | 164(62)      |
| Pulmonary valve stenosis        | 226    | 6                            | 38(3)                  | 99(44)       |
| Mitral valve prolapse           | 63     | 2                            | 90(7.5)                | 44(69)       |
| Aortic valve stenosis           | 35     | 0.9                          | 100(8)                 | 14(40)       |
| PAPVC                           | 17     | 0.5                          | 46 (4)                 | 5(29)        |
| COA                             | 14     | 0.4                          | 80(6.5)                | 7(50)        |

ECD- endocardial cushion defects, VSD- Ventricular septal defect, ASD- Atrial septal defect, PDA- Patent ductus arteriosus, PAPVC- partial anomalous pulmonary connections, COA- coarctation of the aorta

Among the cyanotic defects, Tetralogy of Fallot (TOF) was the most common, 247 cases (7%) were seen with a male preponderance.

**Table 2:** Cyanotic heart diseases

| Lesion                        | Numbers | Mean age Years (months) | Female Number (%) | Overall percentage in CHD (N=3526) |
|-------------------------------|---------|-------------------------|-------------------|------------------------------------|
| Tetralogy of Fallot           | 247     | 4(50)                   | 110(44)           | 7                                  |
| Truncus arteriosus            | 165     | 0.4 (5)                 | 92(56)            | 5                                  |
| DORV                          | 104     | 1.4(16.5)               | 56(53)            | 3                                  |
| Pulmonary atresia             | 71      | 2.6(32)                 | 38(53)            | 2                                  |
| Tricuspid atresia             | 62      | 1.6(20)                 | 31(50)            | 1.8                                |
| D TGA                         | 53      | 0.7(9.5)                | 21 (40)           | 1.5                                |
| A PVR                         | 8       | 0.5(7)                  | 4(50)             | 0.2                                |

D TGA- Transposition of the Great Arteries, APVR- anomalous pulmonary venous return, DORV- Double outlet right ventricle
In addition, sub types of DORV
DORV was diagnosed in 104 children. DORV with sub pulmonic VSD, also called the Taussig Bing anomaly, occurred in 43 children (41%). DORV with sub aortic VSD (VSD physiology), 38 (37%), DORV/ pulmonary stenosis (Fallot type) 15 (14%), and DORV with doubly committed VSD found in 3 (3%). Two (2%) children had DORV with corrected TGA.

| Defect                        | Total number | Female Number (%) |
|-------------------------------|--------------|-------------------|
| Dextrocardia with left isomerism | 14           | 10 (71.4)         |
| Hypoplastic left heart syndrome (HLHS) | 10           | 7 (70)            |
| Ebstein’s anomaly             | 7            | 4 (57)            |
| ALCAPA                        | 3            | 1 (33)            |
| Univentricular heart          | 12           | 6 (50)            |

ALCAPA- Anomalous origin of the left coronary artery from the pulmonary artery

Less common CHDs
Some congenital defects were rare, namely: Aortopulmonary window, which was present in 6 (0.2%) of all CHD. Common atrium 13 (0.3%) with a female preponderance 10 (76%). Partial Anomalous Pulmonary venous connections (PAPVC) were present in 17 (0.5%). It commonly occurred with the sinus venous ASD. Median odiagnosis was 3 years (range 1 month-12 years). Bicuspid aortic valve occurred in 7(0.2%) of all congenital heart disease.

Total anomalous pulmonary venous connections (table 2) were reported in only 8 children (3%); 2 had the infracardiac type, 4 cardiac type and 2 the supra cardiac type.

Patients with dysmorphic features
One hundred eighty-five children had dysmorphic features. The majority 143 (76%), had a phenotypic diagnosis of Trisomy 21 (Down syndrome) with endocardial cushion defects as the most likely diagnosis. Sixty-one of the 186 children underwent genetic testing. Congenital Rubella Syndrome was present in 15 (8) % of cases from data extracted from the WHO-UHI-CRS Surveillance in 2014. Eighty-eight percent had CHD, 68% had ocular defects (cataracts) and 20% had hearing problems. PDA was the most common CHD (77%).
Table 4: Genetic abnormalities

| Genetic disorder                  | N=61 |
|-----------------------------------|------|
| Trisomy 21                        | 24   |
| 22q11.2 deletion syndrome         | 10   |
| Noonan syndrome                   | 5    |
| Turner syndrome                   | 3    |
| Kabuki                            | 2    |
| Holt Oram                         | 1    |
| CHARGE                            | 1    |
| Unrecognized dysmorphism          | 15   |

Of the children with 22q11.2 deletion syndrome, 7 had Tetralogy of Fallot, 2 Truncus Arteriosus and 1 had D-TGA. Noonan's syndrome was confirmed in 5 children; 4 had pulmonary stenosis.

**Congenital heart defects and age at diagnosis**

Most complex heart diseases such as univentricular heart defects, D-TGA, pulmonary atresia, tricuspid atresia, total anomalous pulmonary venous connections and Ebstein's anomaly were rarely diagnosed after the first year of life. Notably, no children with right isomerism/ heterotaxy were seen during the study period.

Table 5: Congenital heart abnormalities and age at diagnosis

| Abnormality                  | Infancy | (>1 < 5 years) | >5 <18Years |
|------------------------------|---------|----------------|-------------|
| VSD                          | 615     | 190            | 116         |
| PDA                          | 556     | 138            | 66          |
| ASD                          | 134     | 114            | 84          |
| Pulmonary Stenosis           | 130     | 69             | 54          |
| ECD                          | 165     | 45             | 55          |
| COA                          | 6       | 2              | 6           |
| TOF                          | 83      | 98             | 66          |
| Truncus Arteriosus           | 144*    | 20             | 1           |
| DORV                         | 72      | 18             | 8           |

*Diagnosed before 6 months of age
VSD, ASD, PDA, TOF, pulmonary stenosis and coarctation of the aorta continued to present in children older than 5 years. Defects not commonly diagnosed after 6 months included; Truncus arteriosus, DORV, Tricuspid atresia, TGA, anomalous pulmonary venous return and Hypoplastic left heart syndrome.

Discussion
This was a retrospective study with a large number of patients making it representative of the entire country. Ventricular septal defects (VSD) were the most common congenital heart defects (26%) with the membranous type in high frequency. Our findings are similar to studies reported elsewhere. Ekure and colleagues reported VSDs in 25% of Nigerian children however, a higher prevalence stated in some studies notably Cameroon, included adults attesting to the fact that VSD patients survive into adulthood. VSD is one of those defects that were diagnosed till 18 years.

Patent Ductus Arteriosus was the second most common defect. This may be due to increased number of premature deliveries, genetic syndromes, maternal rubella infection and peripartum hypoxia. PDA is highly prevalent in extreme preterm babies with birth weight less than 1kg. Premature deliveries in Uganda directly eminent from multiple factors including; low and late antenatal attendance for the recommended visits hence mothers tend to miss drugs like Fansidar that are prophylactic for malaria. Premature deliveries have been linked to placental malaria in some studies, poor maternal preconception nutrition and adolescent pregnancies as well as child spacing less 24 months. Anemia and gestational hypertension are the highest risk factors for preterm deliveries.

PDA is one of the cardiac manifestations of Congenital Rubella syndrome reported in infants whose mothers suffered Rubella infection during pregnancy. Other abnormalities include; VSD, peripheral pulmonary branch stenosis, ocular complications and central nervous system problems. A recently concluded World Health Organization Congenital Rubella Surveillance in Uganda showed a high percentage (77%) of PDA in those infants with serologically confirmed Congenital Rubella Syndrome.

At present, Uganda lacks a National rubella vaccination program, developed countries vaccinate children with the MMR (Mumps Measles Rubella vaccine which drastically dropped CRS incidence). Atrial septal defects ranked third. As reported in other studies there was a female preponderance at 56%, with 88% secundum type. ASDs tend to be well tolerated through infancy and childhood and are still diagnosed into adulthood.

We postulate that very few children of neonatal coarctation were seen because they are not referred early to our center and could have been missed by the primary health care provider. Critical neonatal coarctation often presents as an emergency with a new born in shock and fatal without immediate intervention. Similarly, aortic coarctation among older children was rare. This may imply a low prevalence of this condition or show that man of these patients are not detected because blood pressure measurements are not routinely carried out in children. The few cases in our study presented after 5 years of age. It has also reported in the Nigerian Congenital Heart Disease registry that coarctation of the aorta was one of the rare CHD.

Tetralogy of Fallot remains the most common cyanotic heart defect as has been reported elsewhere. There is a relatively large population of unrepaired patients alive which implies greater survival in less severe cases. This trend has improved with more patients accessing corrective surgery that is now available at the Uganda Heart Institute. By 2014, 80% of the open heart surgeries were performed by our local team and only 20% were referred abroad who mainly included complex congenital heart defects.

Five percent of the patients had Truncus arteriosus which is higher than what is reported in other settings that give an overall prevalence of 2.4%. This was reflected consistently in the number of cases detected on a yearly basis over the study period. Most cases were diagnosed early (before 6 months) owing to an early presentation with heart failure. No new cases were seen in children above 5 years. Truncus arteriosus is associated with a high prevalence of genetic disorders. Thirty-nine percent of the children who underwent genetic testing had truncus arteriosus. This strongly suggests a genetic etiology in our population.

Cases of D-TGA were rare in the study. Transposition of the great arteries has been associated with a high mortality as reported in some studies. The advent of palliative atrial septostomy at the Uganda Heart Institute that acts as a bridge to surgery, offers hope to these critically ill infants who may present with TGA with restrictive interatrial shunts.

Other complex defects were most prevalent in the first year of life. These were not diagnosed after the first birthday. They have been associated with a high mor-
tality, two thirds of children with complex heart defects such as Hypoplastic left/right heart syndrome do not celebrate their first birth day. Unfortunately, limited treatment options are available in the country for such children.

We noted that some CHDs were rare in our study population, a case in point TAPVC (3%) prevalence was comparable to that reported in the Nigerian Congenital Heart Registry (12). Others; Aortopulmonary window, Ebstein’s anomaly and bicuspid aortic valve.

Genetic studies though limited, had a high likely hood of a positive result indicating a need for routine genetic screening in children with congenital heart disease. Deletion 22q11.2 which is associated with immunodeficiency, hypocalcaemia and learning difficulties was also been diagnosed by our team based on clinical findings. Prior knowledge of a genetic syndrome improves surgical outcomes for patients, given the fact that the surgical teams plan for any related complications for such abnormalities. Doell and friends in Switzerland reported no difference between children with genetic syndromes versus those without who underwent open heart surgery for CHD, a genetic syndrome was an independent risk factor for re intubation, and kidney injury.

Digital archiving enabled cases to be discussed with colleagues from other centers and there was an opportunity for re-evaluation of cases at follow up which improved diagnostic accuracy.

Our major limitation was having a retrospective study at a single site whose results may not be fully representative of the nation. However, two other sites have been established in the northern and western parts of the country.

Conclusion

Congenital heart disease is common among children. VSD, PDA and ASD were the commonest acyanotic heart defects while Tetralogy of Fallot and Truncus arteriosus topped the cyanotic defects. Genetic studies are called for in our population to further understand this high prevalence of Truncus arteriosus.

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References

1. Suzanne M. Gilboa, Jason L. Salemi, Wendy N. Nem-bhard, David E. Fixler, Adolfo Correa. Mortality Resulting from Congenital Heart Disease Among Children and Adults in the United States, 1999 to 2006. Circulation. 2010; 122: 2254-2263 PubMed

2. Charlotte Ferencz JR, Robert Mccarter. Congenital Heart Disease: prevalence at live birth, The Baltimore-Washington infant study. American Journal of Epidemiology. 1985;121(1):31-6.

3. Eloi Marijon AT, Sébastian Voicu et al. Prevalence of congenital heart disease in schoolchildren of sub-Saharan Africa, Mozambique 2006 Volume 113, Issue 3, Pages 440–441

4. Tantchou Tchoumi J C, Giamberti A, Ambassa JC, Sadeu JC. Occurrence and pattern of congenital heart diseases in a rural area of sub-Saharan Africa. Cardiovascular Journal of Africa. 2010;21(00).

5. Thakur JS NP, Ahluwalia SK, et al. Integrated community-based screening for cardiovascular diseases of childhood. World Health Forum. 1997;18(1):24-7. PubMed

6. Tantchou T J, Kingue S, Giamberti A, Cirri S, Frigiola A, Butera A. Occurrence, aetiology and challenges in the management of congestive heart failure in sub-Saharan Africa: Experience of the Cardiac Centre in Shisong, Cameroon. Pan African Medical Journal. 2011; Vol 8(1).

7. Leo Lopez, Steven D. Colan, Peter C. Frommelt, Gregory J. Ensing, Kathleen Kendall, Adel K. Youngszai, Wyman W. Lai, and Tal Geva, Recommendations for Quantification Methods During the Performance of a Pediatric Echocardiogram: Journal American Society Echocardiography 2010;23:465-95.

8. http://www.uvri.go.ug/news/uvri-hiv-reference-laboratory-receives-accreditation-college-american-pathologists-28th August 2019.

9. Lars Erik Carlgren. The incidence of congenital heart disease in children born in Gothenburg 1941-1950. Department of Pediatrics, University of Gothenburg, Sweden (1958).

10. Asuquo U. Anita Congenital heart disease in Nigeria Clinical and necropsy study of 260 cases. Archives of Disease in Childhood, 1974, 49, 36.

11. M. J. McLaren, A. S. Lachman, and J. B. Barlow: Prevalence of congenital heart disease in black school children of Soweto, Johannesburg, British Heart Journal, 1979, 41, 554 558
12. Ekanem N. Ekure, Fidelia Bode-Thomas, Wilson E. Sadoh, Adeola A. Otogade et al. Congenital Heart Defects in Nigerian Children: Preliminary Data from the National Pediatric Cardiac Registry. *World Journal for Pediatric and Congenital Heart Surgery* 2017; Vol. 8(6) 699-706

13. Sadoh WE UC, Danies Q. Congenital heart diseases in Nigerian Children: a multicenter echocardiography study. *World Journal of Pediatric Heart Surgery* 2013;4(2):172-6

14. Maria Gillam-Krakauer and Jeff Reese. Diagnosis and Management of Patent Ductus Arteriosus. *Neoreviews*. 2018; 19(7): e394–e402.

15. http://www.everypreemie.org/wp-content/uploads/2016/02/Uganda.pdf(18/october 2017)

16. Malachi Ochieng Arunda, Anette Agardh and Benedict Oppong Asamoah. Survival of low birthweight neonates in Uganda: analysis of progress between 1995 and 2011. *BMC Pregnancy and Childbirth* (2018) 18:189

17. F.T. Cutts, S.E. Robertson, JL. Diaz-Ortega, R. Samuel: Control of rubella and congenital rubella syndrome (CRS) in developing countries, part 1: burden of disease from CRS, *Bulletin of the World Health Organization*, 1997, 75 (1): 55-68.

18. Fatemeh Vaziri, Shaha Roodpeyma, Manuchehr Hekmat. Cardiovascular Malformations in Congenital Rubella Syndrome: A Case Report, *The Iranian Journal of Cardiac Surgery*. 2011, May 60-62.

19. Barnabas Bakamutumaho, Judith Namuyonga, Mathew Cummings, James Eliku, Peter Lwabi, Sulaiman Lubega et al. Congenital rubella syndrome and associated chronic childhood disability in Uganda: implications for control, prevention, and vaccine policy -unpublished data.

20. http://health.go.ug/programs/uganda-national-expanded-program-immunisation-unepiUNEPI Immunization guidelines.

21. Ing FF, Stac TJ, Griffiths SP, Gersony WM: Early diagnosis of coarctation of the aorta in children: a continuing dilemma, *Pediatrics*. 1996 Sep;98(3 Pt 1) 378-82

22. Maureen A. Strafford, Sylvia P. Griffiths, Welton M. Gersony: Coarctation of the Aorta: A Study in Delayed Detection, *Pediatrics* 1982, February vol 62, 2

23. Aliku T, Lubega S, Namuyonga J, Mwambu T, Lwabi P, Sable C: Pediatric Cardiovascular Care in Uganda: Current status, Challenges and opportunities for the future *Annals of Paediatric Cardiology*. *Ann Pediatric Cardiology*. 2017 Jan-Apr; 10(1): 505.

24. Smitha R, Karat SC, Narayappa D et al. Prevalence of Congenital Heart Diseases in Mysore. *Indian Journal of Human Genetics* 2016;12(1):11-16

25. Barakat Adeola Animasahun, Akpoembele Deborah Madise-Wobo, Samuel I Omokhodion, Olisamedua Fidelis Njokanma. Children with Tetralogy of Fallot in an Urban Centre in Africa: *J Cardiovasc Thoracic Res*, 2015, 7(4), 168-171

26. Calder R, Van Praagh R, Van Praagh S, et al. Truncus arteriosus communis. Clinical, angiographic, and pathologic findings. *Am Heart J* 1976; 92(1): 23-38.

27. Barakat Adeola Animasahun, Aminat Titilayo Ogunlana, and Henry Olusegun Gbelee. The Burden of Truncus Arteriosus in an Urban City in Africa: How are we Fairing? *Heart Views*. 2017;18(4): 121–124

28. Doell C, Bernet V, Morinari L Et al. Children with genetic disorders undergoing open-heart surgery: are they at increased risk for postoperative complications? *Pediatr Crit Care Med*. 2011 Sep;12(5):539-44