Profile of Congenital Heart Disease and Access to Definitive Care Among Children Seen at Gulu Regional Referral Hospital in Northern Uganda: a Four-year Experience

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Abstract

Objectives: The aim of this study was to describe the profile of Congenital Heart Disease [CHD] and access to definitive surgical or catheter-based care among children attending a regional referral hospital in Northern Uganda.

Methods: This was a retrospective chart review of all children aged less than 17 years attending Gulu Regional Referral Hospital Cardiac clinic from November 2013 to July 2017.

Results: A total of 299 children were diagnosed with CHD during the period. The median age at diagnosis was 12 months ([QR: 4 – 48] with females representing 59.2% [n=177] of cases. Neonates comprised only 7.4% [n=22]. The commonest CHD seen was ventricular septal defect [VSD] in 19.4% [n=58] of cases, followed by atrioventricular septal defect (AVSD) in 17.1% [n=51] and patent ductus arteriosus (PDA) in 15.7% [n=47]. The commonest cyanotic CHD seen was tetralogy of Fallot [TOF] in 5% [n=15], followed by double outlet right ventricle [DORV] in 4% [n=12] and truncus arteriosus in 3.4% [n=10]. Dextro-transposition of the great arteries [D-TGA] was seen in 1.3% [n=4]. At initial evaluation, 75% [n=224] of all CHD needed definitive intervention and 14% of these children [n=32] had accessed surgical or catheter-based therapy within 2 years of diagnosis. Three quarters of the cases who had intervention [n=24] had definitive care at the UHI including all 12 cases who underwent catheter-based interventions.

Conclusions: There is delayed diagnosis of most rural Ugandan Children with CHD and access to definitive care is severely limited. In-country programs offer the most feasible option to increase access to definitive care.

Background

Globally there has been considerable progress in child survival, and millions of children born today have better survival than in 1990, with world wide mortality rates falling to 39 per 1,000 live births in 2017 compared to 93 per 1,000 live births in 1990 [1] and an estimated annual reduction rate of 4.2% in sub-saharan Africa between 2000–2017 [1]. This has largely been due to considerable investments in public health interventions targeting common childhood illnesses such as vaccination programs, treatment of infectious diseases, and ensuring access to clean water and sanitation among others [1].

However, the story for children born with congenital heart disease [CHD] is different. As preventable communicable diseases decline, there is unmasking of the high proportionate burden of CHD as a contributor to infant mortality. With rising populations and high total fertility rates among women in sub-Saharan African countries, there are rising numbers of children born and or living with CHD in these countries. Lower levels of literacy, poor governance and inadequate financial and skilled human resources for health pose considerable challenges in the implementation of possible interventions to reduce the burden of CHD.

Without early recognition, definitive diagnosis and treatment, it estimated that from one-third to more than half of all children born with significant CHD in developing countries will die in the first month of life [2], while the surviving half will die before their first birth day, making early diagnosis of critical importance. Unfortunately, there are generally few sub-specialist tertiary level pediatric cardiac care services in developing countries, and when available in a country these are often located in the big cities. As a result, many descriptive studies on the burden of CHD in sub-Saharan African countries is limited to data obtained from larger tertiary level health facilities. Furthermore, because of these inequalities in access to pediatric care at regional and district levels, many more rural children with heart disease suffer death or disability compared to those living near cities, often presenting late at initial diagnosis with complications such as failure to thrive heart failure, infective endocarditis and pulmonary arterial hypertension [2, 3].

The aim of this study was to describe the profile of CHD and access to definitive surgical or catheter-based care among children attending a regional referral hospital in Northern Uganda.

Methods

In September 2013 a satellite cardiac clinic was established at Gulu Regional Referral Hospital (GRRH) in Gulu, in northern Uganda [see Fig. 1]. This is a semi-urban town located 335 km from the capital Kampala, serving as a referral point for patients with heart disease in the entire Northern Region of the country that was ravaged by a two-decade civil war. The clinic was open for patient consultations for three working days each week and was staffed with a paediatric cardiologist and two nurses. While this satellite clinic is the only facility providing advanced echocardiographic assessments in the region, a sonographer with general ultrasound skills at the neighbouring St Mary's Hospital, Lacor, located about three miles from GRRH also performs transthoracic echos [TTE] and refers children with suspected heart disease to the cardiac clinic at GRRH.

This was a retrospective chart review. Clinical notes and echocardiographic records of all children diagnosed with CHD aged less than 17 years attending GRRH Cardiac clinic from November 2013 to July 2017 were reviewed. All TTE were performed by a pediatric cardiologist using a portable Vivid Q echocardiography machine [GE Ultrasound, Milwaukee, Wisconsin] Complex and difficult cases were discussed with other pediatric cardiologists at the Uganda Heart Institute [UHI] located in the capital Kampala or from Children's National Health System [USA]. Need for definitive surgery or cardiac catheterization was assessed using established consensus guidelines [4]. Definitive surgical or catheter-based care was either performed at the UHI [the only tertiary facility offering advanced cardiovascular care in the country] or abroad.

Results

A total of 888 children attended the cardiac clinic during the study period [see Fig. 2]. Of these, 33.7% [n = 299] had CHD, whereas 36.1% were normal [n = 321] and 30.2% [n = 268] had acquired heart disease

[RHD, n = 220 and DCMP, n = 22]. The majority of children with rheumatic heart disease [RHD] were diagnosed through a school-based screening program [5]. Females represented 59.2% [n = 177] of cases with CHD. The median age at diagnosis for patients with CHD was 12 months [QR: 4.0–48]. The median age of cases with Acyanotic CHD was 16 months [IQR:5–60] while that of cases with cyanotic CHD was 10 months [IQR:3.5–43]. Neonates [children aged < 28 days] comprised only 7.4% [n = 22] of cases, with infants [children aged 1-12months] representing most cases at 54.6% [n = 125], [see Table 1]. None of the cases had prior prenatal diagnosis.

Table 1
Baseline Characteristics in patients with CHD, N = 299

Variable	**Number (%)**
Gender, Female	177(59.2%)
Median age at diagnosis (IQR), months	12 (4–48)
Age Category at initial diagnosis	
Neonates (0–28 days)	22(7.4%)
Infants (1–12 months)	125(41.8%)
Young child (1–5 years)	83 (27.8%)
Older Child (6–17 years)	69(23.0%)
CHD Type	
Acyanotic CHD	242(80.9%)
Cyanotic CHD	57(19.1%)
The median age of cases with Acyanotic CHD was 16 months (IQR:5–60)	
The median age of cases with cyanotic CHD was 10 months (IQR:3.5–43)	

Acyanotic CHD comprised of 80.9% of all cases of CHD. The commonest CHD seen was ventricular septal defect [VSD] in 19.4% [n = 58] of cases [see Table 2], followed by atrioventricular septal defect [AVSD] in 17.1% [n = 51] and patent ductus arteriosus (PDA) in 15.7% [n = 47]. Complete AVSD was the commonest subtype of AVSD seen [complete, n = 45; transitional, n = 1 and partial, n = 5]. The clinical phenotype of Down syndrome was identified in 46 of cases with AVSD all of whom had complete AVSD. Isolated atrial septal defects [ASDs] were seen in 10% of cases [n = 30] and a patent foramen ovale [PFO] seen in 9.2% [n = 21]. Obstructive lesions were rarely seen, with pulmonic stenosis seen in 3.7% [n = 11] and coarctation of the aorta in 1% [n = 3].

Table 2
Distribution of Acyanotic CHD, n = 242

Type of Lesion	N (Percentage of Total CHD)
Ventricular septal defect (VSD)	58 (19.4%)
Patent Ductus Arteriosus (PDA)	47 (15.7%)
Atrial septal defects (ASD)	30 (10.0%)
Atrioventricular septal defects (AVSD)	51 (17.1%)
Pulmonary stenosis (PS)	9 (3.7%)
Coarctation of Aorta (COA)	3 (1.0%)
Others	47 (15.7%)
Note: Others included defects such as Small Patent Foramen ovale (n = 22), bicuspid aortic valve (4), Isolated persistent LSVC (4 cases), among other defects	

Table 3
Distribution of cyanotic CHD, n = 57

Type of Lesion	N (Percentage of Total CHD)
Tetralogy of Fallot (TOF)	15 (5.0%)
Double Outlet RV (DORV)	12 (4.0%)
Truncus arteriosus (TA)	10 (3.4%)
d-Transposition of the Great Arteries (d-TGA)	4 (1.3%)
Tricuspid Atresia	5(1.7%)
Pulmonary atresia (With one case having intact ventricular septum	5(1.7%)
Others	6 (2.0%)
Others included Double inlet left ventricle (n = 2), Hypoplastic Left heart syndrome (n = 1), Ebstein's anomaly (n = 1), A univentricular heart of RV morphology with pulmonary atresia (n = 1) and a case of left atrial isomerism with common atrium, double out RV and malposed great arteries	

The commonest cyanotic heart diseases seen was tetralogy of Fallot [TOF] in 5% [n = 15] of CHD cases, followed by double outlet right ventricle [DORV] in 4% [n = 12] and truncus arteriosus (TA) in 3.4% [n = 10]. Cases of dextro-transposition of the great Arteries (d-TGA) was seen in 1.3% [n = 4]. During the study only 1% [n = 3] with critical duct dependent lesions were seen [one each with D-TGA with intact ventricular septum, pulmonary atresia with intact ventricular septum and hypoplastic left heart syndrome].

At initial evaluation, 75% [n = 224] of all CHD cases needed definitive intervention, whereas 14% [n = 40] were stable for observation and follow up to determine long-term care plan [see Fig. 3]. Unfortunately, 1% of cases [n = 3] were considered inoperable at initial evaluation [a ten-year-old child with VSD and Eisenmenger syndrome, and two older children having heterotaxy syndrome, complex CHD and severe pulmonary arterial hypertension]. During the study period, 14% of children [n = 32] in need of definitive intervention had accessed surgical or catheter-based therapy. Three quarters of the cases who had access to surgical or transcatheter therapy [n = 24] had definitive care at the UHI including all 12 cases who underwent catheter-based interventions [PDA device occlusion/coil embolization, n = 11 and balloon pulmonary valvuloplasty n = 1]. The catheterization team at the UHI performed all the interventions independently. The cases who underwent surgical care at the UHI included lower risk procedures such as ASD closure [n = 5], PDA ligation [n = 4], coarctation of aorta repair [n = 1], VSD closure [n = 1] and TOF repair [n = 1]. The surgical team at the UHI independently operated on all cases done locally except for 3 ASD cases which were done during surgical camps organized with visiting surgical teams. The local teams combined (both catheterization and surgical) independently performed 87.5% [n = 21] of the cases done locally. The remaining 8 cases obtained definitive surgical care from abroad [VSD closure = 5, ASD closure = 1 and DORV + PS = 1]. All cases who had definitive surgical or transcatheter care where sponsored by grants from charity organizations, except in one case [coarctation of the aorta repair that was subsidized by the UHI]. In two cases, parents declined surgery or transcatheter therapy when they were offered opportunity despite counselling. These included the case of a 15-year-old athletic girl diagnosed with a large secundum ASD through a school based RHD screening program and another case of a 7-year-old girl with a large PDA whose single mother could not find an adult relative to look after 3 younger siblings during a proposed short hospitalization for PDA device closure.

Discussion

In this retrospective review, we have characterized the profile of CHD in a regional referral hospital in Northern Uganda. We found evidence of delayed access to initial diagnosis for CHD, with only 7.4% of patients diagnosed in the neonatal period. The median age at diagnosis of 12 months was substantially longer than the mean age of 5 months reported by Namuyonga et al [6] seen in the Capital Kampala and likely reflects poor screening at lower level health facilities and weak health system referral infrastructure within the catchment area of GRRH. These findings contrast routine fetal and neonatal diagnosis in developed countries [7] causing a shift in the epidemiological profile of CHD secondary to natural selection.

As demonstrated around the world [6, 8–12], VSDs were the commonest forms of CHD diagnosed in Northern Uganda. The peculiar finding in this study was the high number of cases of AVSD seen, accounting for 17.1%, whereas most studies from Africa report prevalence of AVSD between 5–8% of total CHD [9, 11]. As expected, most patients with AVSD in this study had the clinical phenotype of Down's syndrome [representing 90% or 46/51 cases of AVSD] which is like that found by Chelo et al in Cameroon [9] who reported that 80% of cases of AVSD had Down's syndrome. While the high representation of AVSD in this study is surprising, it could be explained by the possible selection bias where prior

echocardiographic screening in a neighbouring hospital which could have resulted in referral of more obvious CHD.

Additionally, as has been reported elsewhere in the country [13], a relatively high number of children with truncus arteriosus cases were seen in this series accounting for 3.4% [n = 10] of total CHD cases. This may be explained by natural selection, or yet to be determined genetic factors. Similarly, the low numbers of cases in this series with critical disease [such as D-TGA, n = 4 and other duct dependent defects n = 3] which are often diagnosed in the newborn period, are likely attributable to early neonatal death before referral for diagnostic evaluation. In this study only three patients were deemed inoperable at index evaluation.

Timely access to initial diagnosis followed by definitive surgical or catheter based cardiac interventions is critical to optimize survival and quality of life for many children with significant CHD [14, 15]. When available in a developing country, these advanced cardiac interventions remain of limited scope because of financial and human resource constraints. In this study, 75% of children with CHD needed definitive intervention at initial presentation, and only 14% of those were able to access definitive surgical or catheter-based therapy within two years of diagnosis. While the 14% access to definitive therapy for CHD in this study is better than the 9% [49/502] access to definitive surgical care for patients in the Ugandan rheumatic heart disease [RHD] registry (16), this study highlights the huge need for advanced tertiary level cardiac care services for patients with heart disease in Uganda. As demonstrated in this study, where three quarters of definitive procedures were performed at the UHI [24 of 32 cases who underwent surgical or catheter-based therapy], with 87.5% of all procedures carried out independently by the local team, developing in-country capacity to deliver tertiary level care for children with CHD remains the single most feasible and cost-effective model to increase access to definitive congenital heart disease care in developing countries. Over the past decade the UHI through its partnerships has invested heavily in building local capacity to perform lifesaving procedures in country, with remarkable progress [16, 17, 18].

Conclusions

There is considerable delay in the initial diagnosis of CHD in our series with less than 10% of children receiving neonatal diagnosis. There was an unusually high proportion of AVSD represented in this series that was mainly associated with Down Syndrome. There is unusually high number of cases with truncus arteriosus where as critical duct -dependent lesions rarely seen, probably related to lack of identification and early mortality in the newborn period. There is need to increase newborn screening in peripheral or regional hospitals to increase identification and referral of children with suspected CHD in the neonatal period through measures such as pulse oximetry screening. Increasing local national capacity for in-country repair of the commonest forms of CHD is essential to increase access to definitive care in those needing intervention.

Abbreviations

ASD: Atrial septal defect; CHD: Congenital heart disease; DORV; Double outlet right ventricle; PDA: Patent ductus arteriosus; TOF; Tetralogy of Fallot; TA: Truncus arteriosus; UHI: Uganda Heart Institute; VSD: Ventricular septal defect

Declarations

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Authors' contributions:

TA provided major contributions in concept, review design, data collection, literature review and drafting the manuscript. AS, AD, JK and RA contributed to data collection, clinical care and manuscript review. AB, CS, SL and PL contributed to concept, study design, clinical care of cases and manuscript review. All authors read and approved the final manuscript.

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Availability of data and materials:

The data sets analyzed during the current study are available from the corresponding author upon reasonable request.

Ethics approval:

The study was approved by the Makerere University School of Medicine Research Ethics Committee as part of a school based Rheumatic Heart Disease screening project

Consent for publication:

Not applicable.

Competing interests:

The authors declare that they have no competing interests.

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Figures

Figure 1

Location of Gulu in Northern Uganda Where Study was done. Note: The designations employed and the presentation of the material on this map do not imply the expression of any opinion whatsoever on the part of Research Square concerning the legal status of any country, territory, city or area or of its authorities, or concerning the delimitation of its frontiers or boundaries. This map has been provided by the authors.

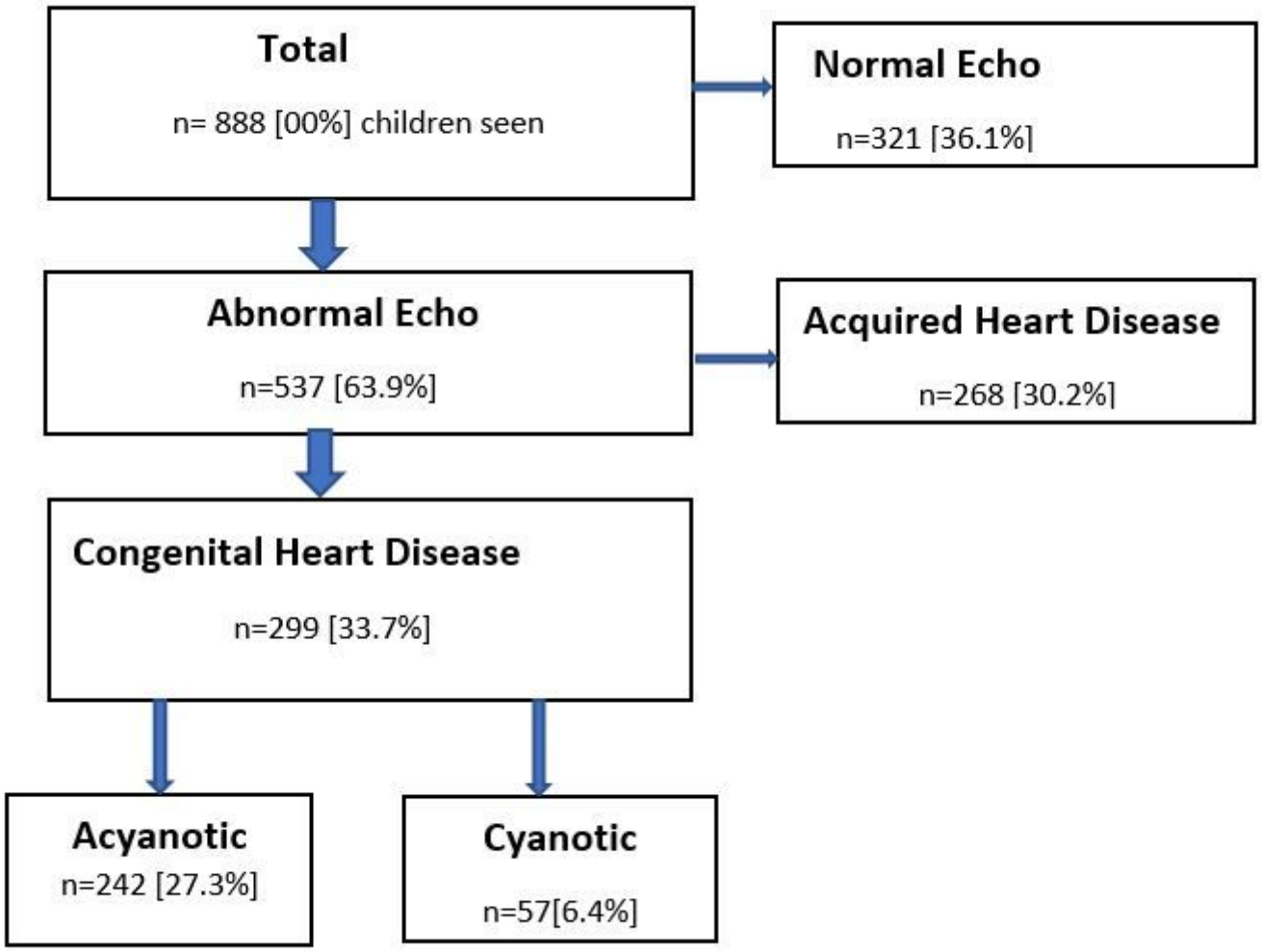

Figure 2

Study Profile Note: Rheumatic Heart Disease [n=220] comprised most cases of acquired heart disease, followed by Dilated cardiomyopathy [n=22].

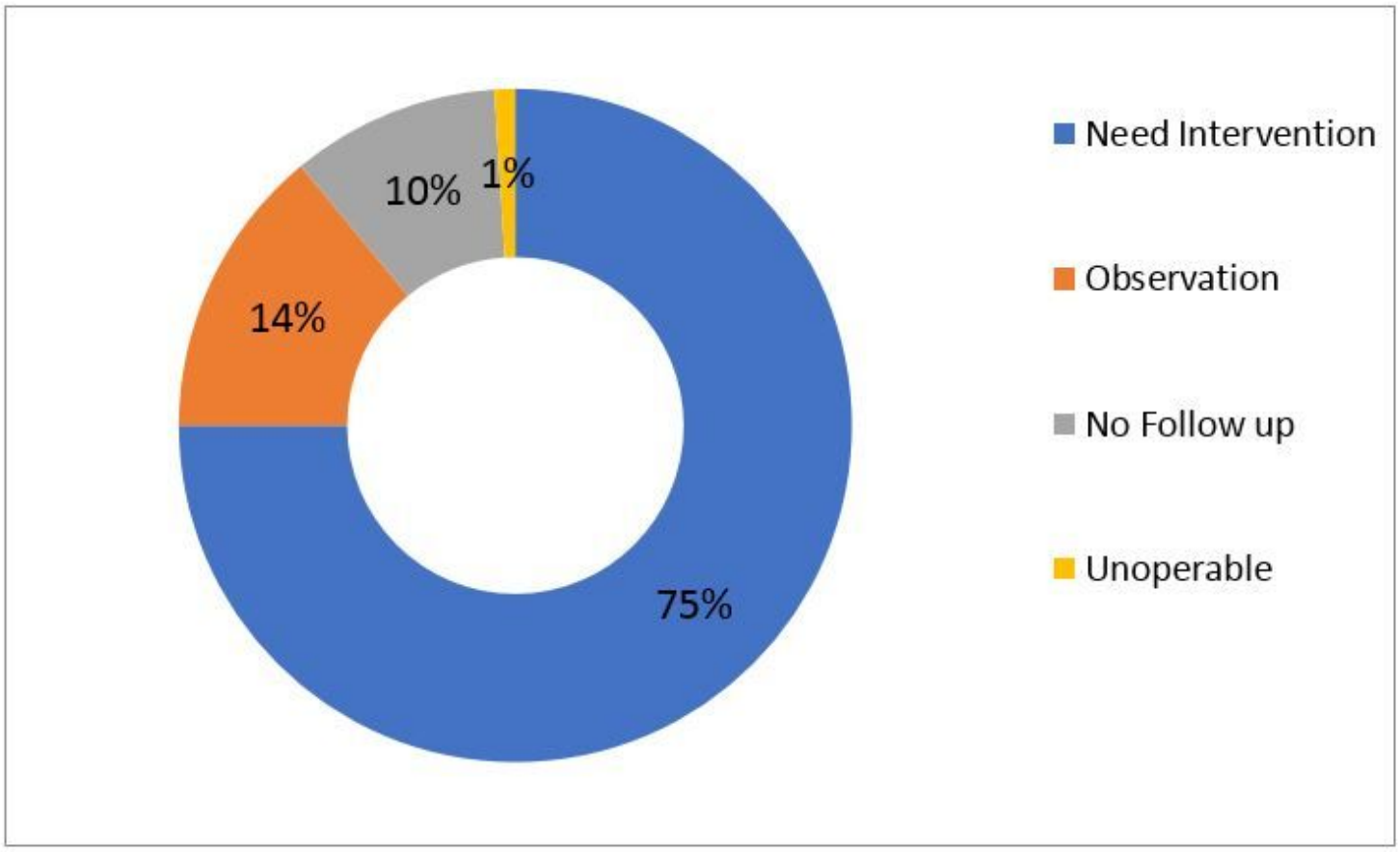

Figure 3

Long term care plan for patients with CHD at initial diagnosis N=299