

# Spectrum of Heart diseases in Children presenting to a Paediatric Cardiac Echocardiography Clinic in the Lake Zone of Tanzania: a 7 years overview

Antke Zuechner (✉ [antke.zuechner@web.de](mailto:antke.zuechner@web.de))

CCBRT <https://orcid.org/0000-0001-5569-276X>

Tumaini Mhada

Bugando Medical Centre

Naizihijwa G Majani

Jakaya Kikwete Cardiac Institute

Godwin G Sharau

Jakaya Kikwete Cardiac Institute

William Mahalu

Bugando Medical Centre

Matthias W Freund

University Hospital Oldenburg

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## Research article

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# Abstract

**Background** Congenital heart diseases (CHD) are among the most common congenital malformations. It is estimated that the incidence of CHD is constant worldwide, but data are rare for most African countries including Tanzania. Even less data are available on the prevalence of acquired heart diseases (AHD) in African children. Rheumatic heart disease is the leading cause of AHD and is remaining a public health concern in Sub-Saharan Africa affecting especially the younger population. Both, CHD and AHD contribute substantially to morbidity and mortality during infancy and childhood.

**Methods** This hospital-based, retrospective review of the registry at the paediatric cardiac clinic of Bugando Medical Centre in the Lake Zone of Tanzania analysed the spectrum of heart diseases of paediatric patients during their first presentation by using simple descriptive statistics.

**Results** Between September 2009 and August 2016, a total of 3982 patients received cardiac evaluation including echocardiography studies. 1830 (46.0%) pathologic findings were described, out of these 1371 (74.9%) patients had CHD, whereas 459 (25.1%) presented with AHD. 53.9% of the patients with CHD were female and the most common associated syndrome was Down syndrome in 12.8% of patients. In 807 patients (58.9%) diagnosis of CHD was established within the first year of life. The majority of patients (60.1%) were in need of surgical or interventional therapy at time of diagnosis and 6.3% of patients were judged inoperable at the time of first presentation. Nearly 50% of cases with AHD were rheumatic heart diseases followed by dilated cardiomyopathy and pulmonary hypertension without underlying CHD.

**Conclusions** The spectrum of CHD and AHD from one centre in Tanzania is comparable to findings reported in other countries from the African continent. Echocardiography is a valuable diagnostic tool and the widespread use of it should be enhanced to diagnose heart diseases in a large number and reasonable time. Most patients present late and majority is in need of surgical or interventional treatment, which is still not readily available. Untreated heart diseases contribute substantially to morbidity and mortality during infancy and childhood. Adequate cardiac services should be established and strengthened.

## Background

Congenital heart diseases (CHD) include a variety of malformations of the heart and major vessels that are present at birth and that are among the most common congenital malformations [1, 2]. It is estimated that the incidence of CHD is constant worldwide [3, 4], though data are rare for most African countries [5, 6]. The estimate of 8 cases per 1000 live births is widely accepted with variations between regions and countries due to genetic and environmental differences [1, 7].

Little is known about the prevalence of CHD in Tanzania and only very few studies have been published [8, 9]. But given a current birth rate of 2.1 million babies per year in the United Republic of Tanzania [10] (UNICEF, 2017) and considering an estimated incidence of 8 cases with CHD / 1000 live births would mean that up to 16.800 infants per year might be born with congenital heart disease.

Even less data are available on the prevalence of acquired heart diseases (AHD) in African children [11]. Rheumatic heart disease (RHD) is still the leading cause of AHD and is remaining a public health concern

in Sub-Saharan Africa affecting especially the young population [11, 12].

Both form of heart diseases together contribute substantially to morbidity and mortality during infancy and childhood in low- and middle-income countries [6, 13]. We therefore report a 7 years overview describing the spectrum of heart diseases diagnosed in a paediatric cardiac echocardiography clinic in the Lake Zone of Tanzania.

## Methods

This hospital-based, retrospective study was conducted at the Bugando Medical Centre (BMC), Mwanza, Tanzania. BMC is one of the four referral hospitals of mainland Tanzania serving for a population of approximately 14 million. The paediatric cardiac echocardiography clinic at BMC is performed twice per week, being the only clinic of its kind in the Lake Zone of Tanzania. Patient data are recorded into a registration book and contain age, gender, type and severity of the heart disease as well as extracardiac malformations like Down syndrome. Furthermore, a conclusion concerning the need for an intervention, e.g. surgery, is documented at the time of presentation. Children below 16 years of age who presented to the clinic for the first time and were diagnosed with an abnormal echocardiography study were included into the review. Cardiac consultation and echocardiography were performed by a paediatric cardiologist and by well-trained and supervised paediatricians (AZ, NM, TM). The echocardiography including two-dimensional-, colour-, pulse wave- and continuous wave-imaging was performed using two portable echocardiography machines, Sonosite M-Turbo and GE Logic i.

The data were entered into Excel and then analysed by using simple descriptive statistics.

Quality control was ensured through visiting international cardiologists during cardiac missions and through intraoperative findings and diagnostic cardiac catheterizations done outside the country.

## Results

Between September 2009 and August 2016, a total of 3982 children underwent cardiac evaluation and echocardiographic examination at the paediatric cardiac clinic of BMC. In 1830 cases (46.0%) pathologic findings were described, out of these 1371 (74.9%) patients had congenital heart diseases (CHD), and 459 (25.1%) presented with acquired heart diseases (AHD).

Figure 1: Distribution of congenital vs acquired heart diseases

## Congenital Heart Diseases

53.9% of the patients with CHD were female and 19.3% of all cardiac patients presented with associated syndromes or extracardiac malformations. The most common associated syndrome was Down syndrome in 12.8% of the patients. Leading cardiac lesion was Ventricular septal defect (VSD) in 358 patients (26.1%) followed by Patent ductus arteriosus (PDA) in 318 patients (23.2%), which included also

51 premature infants admitted to the neonatal department of BMC. Most common cyanotic heart disease was Tetralogy of Fallot (ToF) in 10.1% of cases. The distribution and frequency of CHD are shown in table 1.

208 (15.2%) out of the 1371 patients with CHD were diagnosed in the first month of life, by six months of life diagnosis was made in 42% of patients and close to 60% of patients were at least diagnosed within the first year of life as shown in table 2. Approximately 40% of the children with VSD as the leading CHD were diagnosed within the first 6 months of life, whereas children with ToF as the most common acyanotic CHD were usually diagnosed later in life and only one third were diagnosed within the first year of life. The disease specific age distribution of these two diseases is presented in table 3a/b.

At time of diagnosis approximately 60% of patients (824 / 1371) were judged to be in the need of surgical or interventional treatment and in 87 patients (6.3%) signs of severe pulmonary arterial hypertension (Eisenmenger syndrome) were present. Another 68 patients (5.0%) were in need of diagnostic catheterization to assess eligibility for surgery. The distribution of cases with respect to need of further diagnostic assessment or treatment is shown in table 4.

Seven patients (0,5%) were diagnosed with CHD while they presented to our hospital with acute infective endocarditis or endarteritis and intracardiac and intraarterial vegetations were demonstrated by echocardiography.

Nearly every fifth patient (19.3%) with congenital heart disease showed extracardiac malformations with Down syndrome being the leading associated syndrome. The distribution is depicted in table 5.

## Acquired heart diseases

Almost 50% of cases with acquired heart diseases were rheumatic heart diseases (RHD) followed by dilated cardiomyopathy and pulmonary hypertension without an underlying structural heart disease. Table 6 is presenting the distribution and frequencies of AHD.

The majority of patients diagnosed with RHD presented with isolated mitral valve regurgitation (158 patients / 69.3%), whereas 8.3% showed multivalvular disease followed by isolated aortic valve regurgitation (7.5%) and combined mitral valve regurgitation and mitral valve stenosis (6.6%).

At time of diagnosis most patients presented with advanced RHD and an indication for surgery (valve repair or valve replacement) was given in 197 out of 228 cases (86.4%).

## Discussion

Limitations of the study: This study is a single centre, hospital based review and not a community based study. Therefore, the study does not provide information on the prevalence of heart diseases in the

general paediatric population of Tanzania. Furthermore, it is a retrospective review of the echocardiography registry which might contain some incomplete data or missing information.

**Strengths of the study:** This study comprises a large sample size compared to other studies and patients were examined regardless of their financial status or availability of health insurance.

The overall distribution of heart diseases in congenital and acquired cases corresponds well with the findings of a comparable study in Cameroon, where in 1666 patients with heart diseases, 73.8% of cases presented with CHD and 25.8% with AHD [14].

Assessing the distribution of CHD in our review, VSD was the most common type, which is consistent with the results of two meta-analyses [1, 4] and a literature review [15] for the worldwide prevalence of this heart defect. Though most of the studies done in developing countries demonstrated even a higher prevalence of VSD ranging from 30% to 58% [14, 16, 17, 18] compared to our result of 26.1%.

As characteristic for countries with low resources and limited access to cardiac surgery Tetralogy of Fallot (ToF) is the leading cyanotic heart disease and the frequency is higher than documented in developed countries [14, 17, 18, 19]. Children with ToF do not present with heart failure, but with progressing signs of chronic cyanosis (e.g. finger clubbing) and frequent squatting. Many of them show a naturally balanced pathophysiology and survive without intervention whereas infants suffering from other cyanotic heart diseases might die early and even undiagnosed [17, 20]. This is also supported by our findings of diagnosis specific age distribution among children with VSD and ToF. Forty percent of children born with VSD are diagnosed within the first 6 months of life, whereas children suffering from ToF were usually diagnosed later in life and only one third within the first year of life.

Interestingly, the frequency of AVSD (10.5%) in our study was higher than in any other African study. In general, the frequency of AVSD in studies from developing countries is reported to be between 3.5 and 8.8% [17, 21, 22]. The high prevalence of AVSD, mainly in association with Down syndrome (DS), was a constant finding throughout the consecutive years. The prevalence of DS in Tanzania is not known, but studies from Nigeria and South Africa have stated that the prevalence of DS in Sub Saharan Africa is exceeding the reported prevalence of 1 in 750 live births in western countries [23, 24]. This may be influenced by prenatal diagnosis and termination of pregnancy in the western countries [25]. Furthermore, a recent study from Nigeria examined the prevalence of CHD in children with DS and found a prevalence of 75%, which is significantly higher than reported by others [26].

Overall, the rate of associated extracardiac malformations corresponds well with the findings of other studies [25, 27, 28].

RHD presented as the leading cause of AHD in children in our study similar to what is consistently reported worldwide [13]. It is furthermore very likely that the condition is widely underreported in our review as the vast majority of patients was diagnosed with advanced findings and we didn't detect any subclinical RHD as reported in other reviews [11].

## Conclusion

The spectrum of congenital and acquired heart diseases presenting to the paediatric cardiac echocardiography clinic of BMC is corresponding well to the findings reported in other countries from the African continent.

The majority of children with heart disease presenting to our clinic is in need of surgical or interventional treatment, which is not readily available. Furthermore, diagnosis is often delayed and as a consequence a significant number of patients show severe complications and an advanced stage of the heart disease on initial diagnosis. Untreated heart diseases contribute substantially to morbidity and mortality during infancy and childhood. Adequate local cardiac services should be established and strengthened.

Echocardiography is a valuable diagnostic tool as it is non-invasive and cost-effective. The widespread use of it should be enhanced with more paediatricians being trained in this technique, especially in settings with low resources. In a first step a setting of paediatricians trained by paediatric cardiologists to perform echocardiography in the rural areas with mobile echocardiography equipment under the supervision of a local paediatric cardiac centre has to be the goal to diagnose CHDs and AHDs in a large number and in a reasonable time.

Children diagnosed with CHD should be carefully examined for extracardiac malformations as the association between CHD and extracardiac malformations is significant.

All children with Down Syndrome should routinely receive a cardiac examination because of the high association of CHD. A further study would be needed to determine the prevalence of CHD in children with DS in Tanzania.

Rheumatic heart diseases are certainly underreported as most patients were diagnosed with advanced findings and no cases of subclinical RHD were detected. Further population based prevalence studies are required to determine the real burden of this disease and to guide preventive programmes.

## List Of Abbreviations

CHD: Congenital heart disease; AHD: acquired heart disease; RHD: Rheumatic heart disease; BMC: Bugando Medical Centre; VSD: Ventricular septal defect; PDA: Patent ductus arteriosus; ToF: Tetralogy of Fallot; AVSD: Atrioventricular septal defect; DS: Down Syndrome.

## Declarations

## Ethics approval

The review was conducted after approval by the joint ethical research committee of the Catholic University of Health and Allied Sciences—Bugando with Research Clearance Certificate No

## **Consent for participation and publication**

Not applicable

## **Availability of data**

The datasets analysed during the current study are available from the corresponding author upon reasonable request.

## **Competing interests**

The authors declare that they have no competing interests.

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## **Authors' contributions**

AZ provided major contributions in concept, review design, data collection, literature review and drafting the manuscript. TM contributed to concept, review design and data collection. NM contributed to data collection, literature review and writing the manuscript. GS and WM contributed to data collection and manuscript. MF contributed to concept, literature review and manuscript.

All authors read and approved the final manuscript.

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## **Author's information**

AZ is paediatric cardiologist and neonatologist working at BMC during the study period and served as a senior lecturer at CUHAS.

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## Tables

Table 1: Distribution and relative frequencies of CHD

Diagnosis	Number (N = 1371)	%	Comment
Ventricular septal defect (VSD)	358	26.1	
Patent ductus arteriosus (PDA)	318	23.2	Including 51 PDA in premature neonates (3.7%)
Atrioventricular septal defect (AVSD)	144	10.5	98 in children with Down syndrome
Tetralogy of Fallot (ToF)	138	10.1	16 with pulmonary atresia
Atrial septal defect sec. type (ASDII)	98	7.1	
Pulmonary stenosis (PS)	58	4.2	
Truncus arteriosus	50	3.8	
Double outlet right ventricle (DORV)	47	3.4	
Tricuspid atresia	26	1.9	
Transposition of great arteries (TGA)	25	1.8	15 with large VSD
Complex cyanotic CHD	36	2.6	
Others	73	5.3	

Table 2: Age at diagnosis of CHD

Age at diagnosis	Number (N = 1371)	%	cumulative	%
0-1 month	208	15.2	208	15.2
2-6 months	367	26.8	575	42.0
7-12 months	232	16.9	807	58.9
2-5 years	403	29.4	1210	88.3
> 5 years	138	10.1	1348	98.3
Missing data	23	1.7	1371	100

Table 3a: Age at diagnosis in patients with VSD

Age at diagnosis <b>Patients with VSD</b>	Number (N = 358)	%	cumulative	%
0-1 month	31	8.7	31	8.7
2-6 months	113	31.6	144	40.2
7-12 months	61	17.0	205	57.3
2-5 years	109	30.4	314	87.7
> 5 years	37	10.3	351	98.0
Missing data	7	2.0	358	100

Table 3b: Age at diagnosis in patients with TOF

Age at diagnosis <b>Patients with ToF</b>	Number (N = 138)	%	cumulative	%
0-1 month	9	6.5	9	6.5
2-6 months	18	13.0	27	19.6
7-12 months	17	12.3	44	31.9
2-5 years	65	47.1	109	79.0
> 5 years	26	18.8	135	97.8
Missing data	3	2.2	138	100

Table 4: Need for further assessment or treatment

Condition	Number (N = 1371)	%
Mild CHD without need for surgery	392	28.6
Indication for surgery / intervention given	825	60.1
Diagnostic catheterization needed	68	5.0
Signs of severe pulmonary hypertension	87	6.3

Table 5: extracardiac malformations / syndromes

Condition	Number (N = 1371)	%
Down syndrome	176	12.8
Syndromic features (not classified)	35	2.6
Anorectal malformation	15	1.1
Multiple malformation	14	1.0
Rubella syndrome	12	0.8
Others*	13	0.9
Associated extracardiac malformations / syndromes	265	19.3

\*Single cases of Omphalocele, Marfan Syndrome, Williams Syndrome, Turner Syndrome, connective tissue disease, Trisomy 18, missing sternum, ectopic Heart, and duodenal atresia

Table 6: Distribution and frequencies of AHD

Diagnosis	Number (n = 459 cases)	%	Additional information
Rheumatic Heart disease (RHD)	228	49.7	
Dilated Cardiomyopathy (DCM)	74	16.1	16 cases of HIV
Pulmonary Hypertension (PAH)	48	10.5	4 cases of Schistosomiasis
Pericardial effusion (PE)	33	7.2	10 cases of TB
Other cardiomyopathies	29	6.3	10 cases of malignancies
Persistent pulmonary hypertension of the newborn (PPHN)	27	5.9	
Endomyocard Fibrosis (EMF)	20	4.3	

## Figures

### Type of Heart disease



Figure 1

Distribution of congenital vs acquired heart diseases