ORIGINAL ARTICLE

The Major Clinical Presentations and Echocardiographic Features of Structural Cardiac Disease among Children in Kordofan, Sudan

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ABSTRACT

Background: Patterns of heart diseases differ between the high income countries and low income countries.[1, 2] Acquired heart diseases like RHD, cardiomyopathies, pericarditis are still a public health burden while congenital heart diseases still carry a poor outcome among children[3]. The absence of early diagnosis, surgical intervention and inadequate access to medication and the existing burden of communicable diseases increases the strain on a staggering economy of most sub Saharan countries.

Aim: To assess the clinical and echocardiographic pattern of cardiac disease in children in Kordofan state and to develop a road map plan of their further management.

Methods: An analytical cross sectional hospital based study. The study included the children of age groups 1 day up to 18 years with diagnosed or strongly suspected congenital or acquired cardiac disease who have their follow up in the pediatric sections of El-Obied hospitals in the western central part of the Sudan. Every child in the study group was assessed clinically with focused history taken and relevant physical examination. Chest x-ray and proper echocardiography was done for every child in the study group. Management plan was put by the pediatric cardiologist for all the children in the study group. Eighty children were enrolled in this study.

Results: 80 children studied, 55% were males and females represent 45%. Most of the children with cardiac disease in the study were from the age group 1-5 years (27.5%) followed by infants (26.2%).after that, 5-10 years, more than 10 years and less than one month. Nine patients (11.25%) were severely affected, 26 (32.5%) were moderately affected and the growth of 45 (56.25%) was normal. Clinical evaluation of the study group revealed that, 19 children (24%) presented with respiratory symptoms and /or respiratory distress. A cardiac murmur was heard in most of the patients in this series, systolic murmurs heard in 55 patients (69%) of which 21 were pansystolic (26%) and 8 were ejection systolic murmurs (10%). Hepatomegaly was found in 25 patients (31%) and Hepatosplenomegaly in 4 patients (5%). Down syndrome was found in 5 patients (6%), William syndrome in one patient. Congenital HD found in 56 patients (70%) and acquired HD in 24 (30%), 19 patients (79% of the acquired heart disease and 23.7% of the total study group was diagnosed as having rheumatic heart disease and cardiomyopathy was found in 5 patients (20.8% of acquired heart disease).

Conclusion: Rheumatic cardiac disease is preventable through the proper and complete antibiotic treatment of streptococcal pharyngitis and tonsillitis and it is vital to apply and strict to the recommendations of the national program of prevention of rheumatic fever and rheumatic heart disease.

Keywords: Echocardiography, Rheumatic Cardiac Disease, Management, Children, Sudan

INTRODUCTION

The estimate of 8 per 1000 live births is generally accepted as the most reliable incidence of congenital heart diseases, however a recent systematic review emphasized significant differences, with the lowest prevalence rate observed in Africa, particularly among lowest-income populations. This mainly reflects the paucity of readily available estimates and the urgent need for data to support burden of disease calculations in Africa. The number of children and adults affected with CHD in Africa may be underestimated. Several recent publications have profiled the epidemiology of CHD in children and adults in Africa, emphasizing the burden of CHD among patients referred with suspected heart disease. According to recent data from Sudan, the prevalence appears to be dramatically much higher[8].

A hospital-based registry showed that rheumatic heart disease is clustered in Kordofan, Darfur, and White Nile and to a lesser extent in Al Gazira[9]. Handheld echocardiographic screening of 12,000 subjects in five regions revealed a wide disparity in RHD prevalence ranging from 0.3/1,000 in Khartoum to 61/1,000 in North Kordofan.[10] Recent study done in 2017 concluded that the prevalence of rheumatic heart disease in Kordofan is 61.5/1000[10].

This study was conducted to assess the clinical and echocardiographic pattern of cardiac disease in children in Kordofan state and to develop a road map plan of their further management.

METHODOLOGY

This study is an analytical cross sectional hospital based study. The study included the children of age groups 1 day up to 18 years with diagnosed or strongly suspected congenital or acquired cardiac disease who have their

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follow up in the pediatric sections of El-Obied hospitals in the western central part of the Sudan. El-Obied is the capital city of Kordofan and a major city in the Sudan. Ethical considerations and roles were followed and informed parental consent of parents or caregivers was taken for every child enrolled in this study. Every child in the study group was assessed clinically with focused history taken and relevant physical examination .chest x-ray and proper echocardiography was done for every child in the study group. Management plan was put by the pediatric cardiologist for all the children in the study group. Eighty children were enrolled in this study. SPSS software was used in data entry and analysis.

RESULTS

A total of 80 patients was examined during this study, 44 (55%) were Males and36 (45%) were females.

When assessing the growth of the study group, nine patients (11.25%) were severely affected, 26 (32.5%) were moderately affected and the growth of 45 (56.25%) was normal (Fig. 1).

Clinical evaluation of the study group revealed that, 19 children (24%) presented with respiratory symptoms and /or respiratory distress. Severe respiratory symptoms in 6 patients (7.5%). 18 children presented with various grades of cyanosis (22.5%), 11 children (14%) showed heart failure when examined clinically. A cardiac murmur was heard in most of the patients in this series, systolic murmurs heard in 55 patients (69%) of which 21 were pansystolic (26%) and 8 were ejection systolic murmurs (10%). Predominantly diastolic murmurs in only 4 patients (5%) and continuous murmurs in 5 patients (6%). No murmur was detected in 12 children (15%).

Hepatomegaly was found in 25 patients (31%) and Hepatosplenomegaly in 4 patients (5%). Other clinical associations/complications include infective endocarditis features in 4 patients and atrial fibrillation in 2 patients. Down syndrome was found in 5 patients (6%), William syndrome in one patient. One patient presented with active rheumatic fever, one with rheumatic chorea, one with end stage renal disease on dialysis and one patient with sickle cell anemia.

Congenital HD found in 56 patients (70%) and acquired HD in 24(30%), 19 patients (79% of the acquired heart disease and 23.7% of the total study group) was diagnosed as having rheumatic heart disease and cardiomyopathy was found in 5 Patients (20.8% of acquired heart disease). From the 65 patients diagnosed as having CHD, 20 (35.7%) have cyanotic congenital heart disease, 31(55%) have a cyanotic left to right shunt, and 5 (8.9%) with vascular malformation. 15 patients (78.9%) of those diagnosed as RHD were found to have mitral pathology (mitral regurgitation with or without stenosis), and only 4 have aortic regurgitation.

39 patients (49%) of patients were planned for urgent or early surgery or diagnostic and therapeutic intervention. 9(11%) needed acute medical treatment for HF OR infective endocarditis and Long-term medical treatment and follow up was the plan in 32 patients (40%). Table no. 4, showing pattern of the cardiac diseases significantly (p=0.008) differed according to the gender significant, however male patients were more prone to have cardiac diseases than female patient. And other result shows in table no. 5, revealed that initial presentation of the cardiac illnesses were strongly associated (p=0.000) with the age of the patient .

Table 1: Age distribution children of the study group

Age group	No.	%
< 1month	3	3.8
1mo—1year	21	26.2
1year—5 years	22	27.5
5years---10 years	18	22.5
>10 years	16	20.0
Total	80	100.0

Table 2: Chest X-ray features of the children of the study group

X-ray feature	No.	%
Normal chest x-ray	17	21
Cardiomegaly & pulmonary plethora	13	16
Pulmonary oligemia	8	10
Huge cardiomegaly	5	6.5
Dextrocardia	2	2.5
Cardiomegaly with chamber hypertrophy	35	44

Table 3: The main structural echocardiographic features in the study group according to the main lesion

Echo finding	No.	%age
TGA with a shunt	3	3.75
Tetralogy of fallot	9	11
Rheumatic MR+/-stenosis	15	19
Rheumatic AR	4	5
Large to moderate VSD	18	22.5
ASD	3	3.75
Truncus arteriosus	1	1.5
Tricuspid atresia	2	2.5
Pulm.atresia/steosis+/- VSD	6	7.5
Single ventricle/atrium /complex cyanotic HD	3	3.75
PDA	4	5
PFO	2	2.5
AV canal	2	2.5
cardiomyopathy	5	6
others	3	3.75
total	80	100

Table 4: Pattern of cardiac diseases based on structural echocardiographic features among gender with cardiac illness.

	Male	Female
Tetralogy of fallot	10(12.5%)	2(2.5%)
Rheumatic MR+/- stenosis	5(6.3%)	16(20%)
Rheumatic AR	1(1.3%)	0(0%)
Large tp moderate VSD	12(15%)	8(10%)
ASD	0(0%)	2(2.5%)
Truncusarteriosus	0(0%)	1(1.3%)
Tricuspid astresia	1(1.3%)	0(0%)
Pulm. atresia/steosis +/- VSD	1(1.3%)	0(0%)
Single ventricle/atrium / complec cyanotic HD	2(2.5%)	0(0%)
PDA	3(3.8%)	2(2.5%)
PFO	2(2.5%)	0(0%)
AV canal	1(1.3%)	1(1.3%)
Cardiomyopathy	1(1.3%)	4(5%)
Others	5(6.3%)	0(0%)
Total	44(55%)	36(45%)

P value=0.008

Table 5: Initial presentation/Symptoms according to age of the patients:

	Age (In year)					
	1	2	3	4	5	P-value
Incidental murmur	1(1.3%)	9(11.3%)	10(12.5%)	5(6.3%)	2(2.5%)	
Cyanosis/squatting	0(0%)	2(2.5%)	9(11.3%)	1(1.3%)	0(0%)	
Rheumatic fever	0(0%)	0(0%)	0(0%)	9(11.3%)	8(10%)	
Respiratory symptoms	0(0%)	2(2.5%)	6(7.5%)	0(0%)	1(1.3%)	
Dysmorphism/Down syndrome	0(0%)	3(3.8%)	1(1.3%)	0(0%)	0(0%)	
Growth failure	0(0%)	4(5%)	0(0%)	0(0%)	0(0%)	
Heart failure	0(0%)	1(1.3%)	0(0%)	0(0%)	0(0%)	0.000
Others	2(2.5%)	1(1.3%)	0(0%)	1(1.3%)	2(2.5%)	
Total	3(3.8%)	22(27.5%)	26(32.5%)	16(20%)	13(16.3%)	

Fig. 1: Growth Pattern of children of the study group

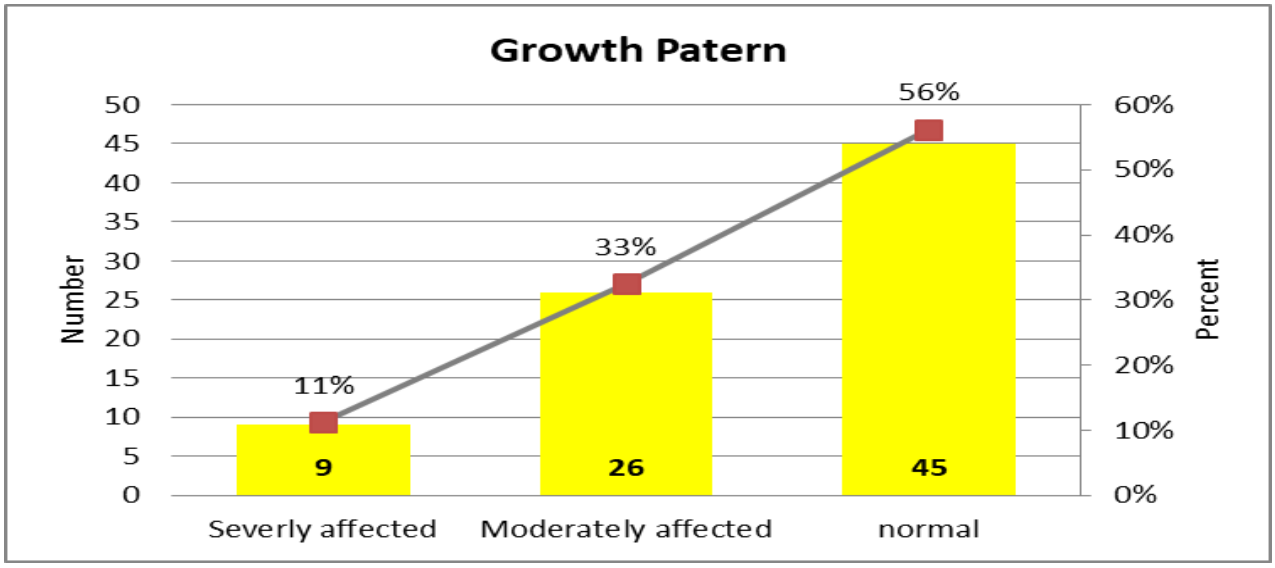


Fig. 2: Initial Presentation/symptoms of children of the study group

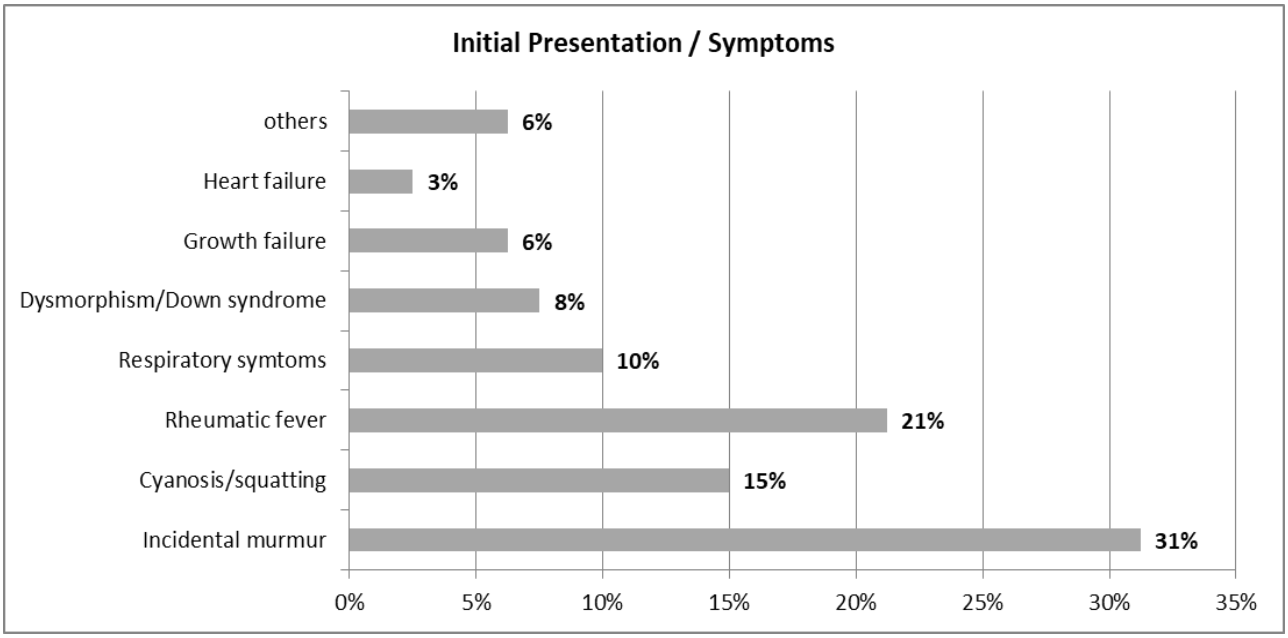


Fig. 3: Chest x-ray features of children of the study group

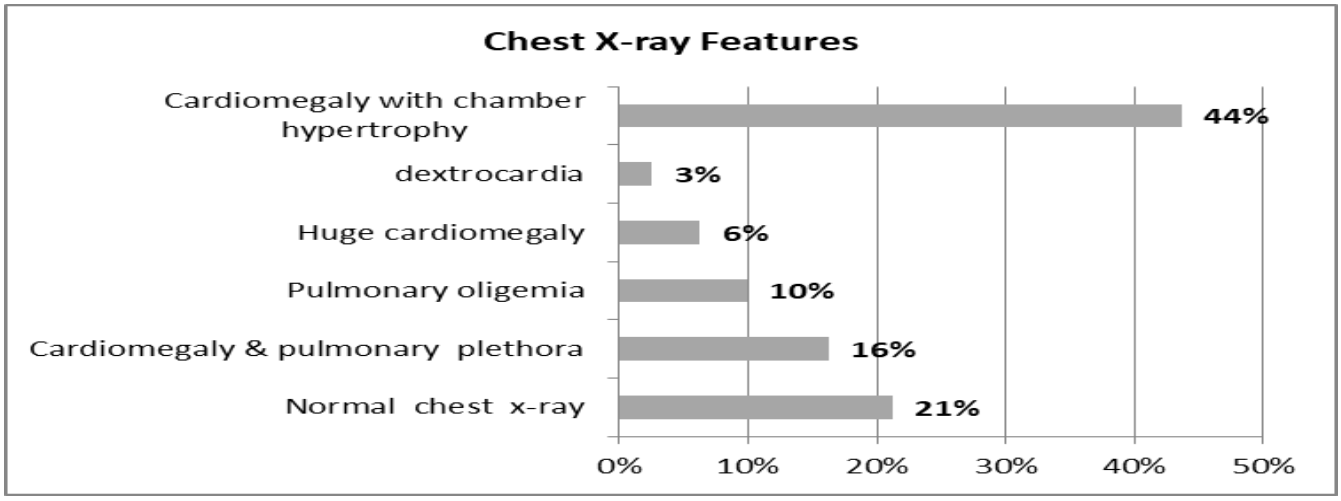


Fig. 4: Main structural echocardiographic features of children of the study group

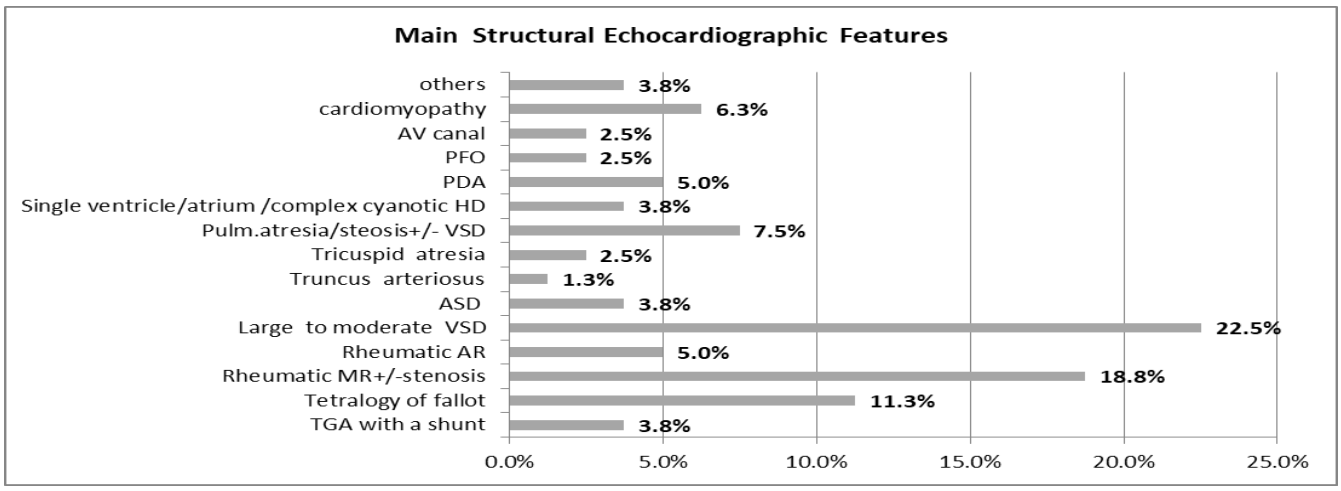
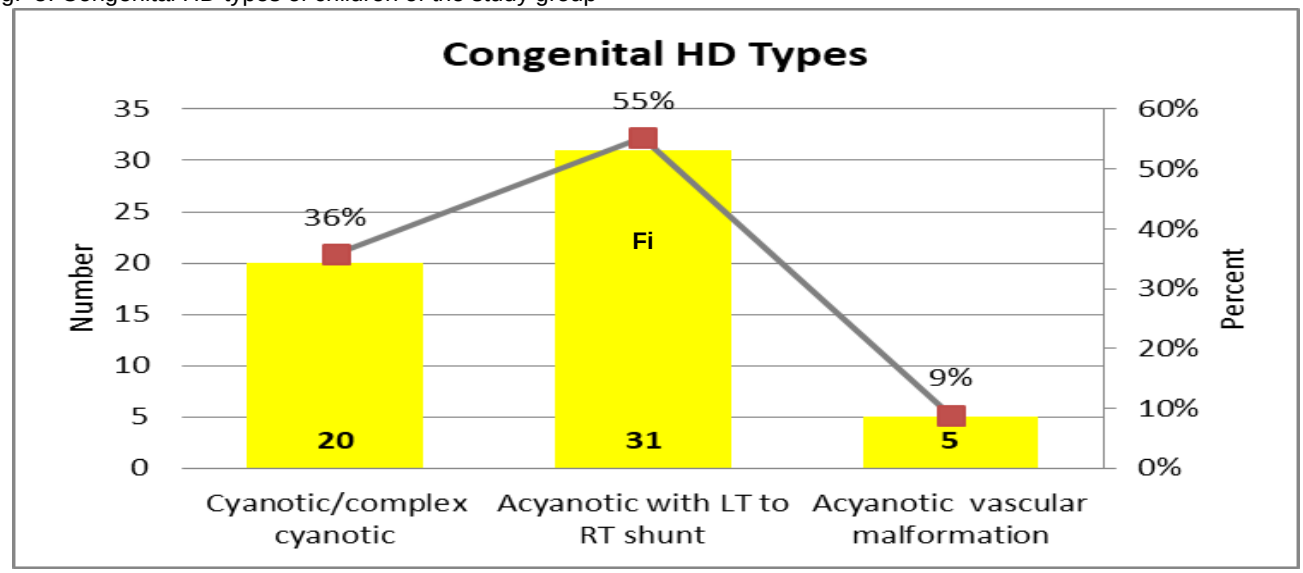


Fig. 5: Congenital HD types of children of the study group



DISCUSSION

From the 80 children studied, 55% were males and females represent 45%. Most of the children with cardiac disease in the study were from the age group 1-5 years (27.5%) followed by infants (26.2%).after that, 5-10 years, more than 10 years and less than one month. Representing 22.5%, 20% and 3% respectively.

When assessing the growth of the study group, nine patients (11.25%) were severely affected, 26 (32.5%) were moderately affected and the growth of 45 (56.25%) was normal. Similarly, in Kenyan children with rheumatic heart disease 50% and 52% of males and females respectively were found having normal weight for age.[11]

Regarding the clinical evaluation, the presenting signs and symptoms were as follows: 19 children (24%) presented with respiratory symptoms and /or respiratory distress. Severe respiratory symptoms in 6 patients (7.5%). 18 children presented with various grades of cyanosis (22.5%). 11children (14%) showed heart failure when examined clinically. The respiratory symptoms were the commonest after the incidental murmur, this was also the case in Indian study[27] and also in Nigeria,[12] the most common presenting complaint among the patients was cough in 17 (43.6%), followed by difficulty in breathing in 12 (30.8%), fever in 10(25.6%) and fast breathing in 8(20.5%).

A cardiac murmur was heard in most of the patients in this series systolic murmurs heard in 55 patients (69%) of which 21 were pansystolic (26%) and 8 were ejection systolic murmurs (10%)predominantly diastolic murmurs in only 4 patients (5%) and continuous murmurs in 5 patients(6%). No murmur was detected in 12 children (15%). Hepatomegaly was seen in 25 patients (31%) Hepatosplenomegaly in 4 patients (5%).Other clinical associations /complications include infective endocarditis features in 4 patients, atrial fibrillation in 2 patients, Down syndrome was found in 5 patients (6%) and William syndrome in one patient.

One patient presented with active rheumatic fever, one with rheumatic chorea, one with end stage renal disease on dialysis and one patient with sickle cell anemia.

Congenital HD found in 56 patients (70%) and acquired HD in 24 (30%), 19 patients (79% of the acquired heart disease and 23.7% of the total study group) was diagnosed as having rheumatic heart disease and cardiomyopathy was found in 5 Patients(20.8% of acquired heart disease). In similar study done at Sudan Heart Centre, Congenital heart disease constituted 87% and acquired heart disease and rhythm disorders 13%.[8] Showing fewer patients with acquired heart disease than the recent study. Another former study at pediatrics cardiac clinic in Khartoum has approximately similar percentages showing that 56% of the patients had congenital heart disease, and 39%had rheumatic heart disease.[13] Recent study done in 2017 concluded that the prevalence of rheumatic heart disease in Kordofan is 61.5/1000.[10] The situation is not different in other African countries, a study done in a cardiology clinic in Addis Ababa, Ethiopia over 20 months showed that over 338 with defined pathology, 152 (44.9%) had rheumatic heart disease.[14] Echocardiographic assessment of 2501 Rwandan school children from 10 schools prevalence was 6.8/1 000 children comparing with the situation in Sudan, RHD prevalence was ranging from 0.3/1,000 in Khartoum to 61/1,000 in North Kordofan[9].

Outside Africa, a study done in Nepal concluded that, from 218 pediatric cardiac cases(66.05%) were Congenital Heart Disease and (26.14%) were Rheumatic Heart Disease, 14 cases (6.42%) were Pericardial Disease and 3 cases (1.37%) were classified as Dilated Cardiomyopathy.

15 patients (78.9%) of those diagnosed as RHD were found to have mitral pathology (mitral regurgitation with or without stenosis), and only 4 have aortic regurgitation. This result is in agreement of the former study done in Kordofan where the mitral regurgitation was the dominant with a percentage of 79.8%, followed by Combined Mitral and Aortic Regurgitation (12%), Aortic regurgitation (3.8%), Combine Mitral stenosis and Regurgitation (2.1%), Mitral Stenosis (1.6%) and lastly combined mitral regurgitation and stenosis and aortic regurgitation 1 (0.7%).[10] Also a study done in 2 hospitals in Khartoum, Sudan concluded that most of the cases (93%) had mitral valve involvement. But a study done in Nigeria showed that the majority of the study population have combined MR and AR[15]. Another Nigerian study concluded that Mitral regurgitation was the commonest echocardiographic diagnosis present in 38%, 27.9% of patients had mixed mitral valve disease (both together 65.9%). This result is closer to our situation than the other study done in the same country.[16] In addition a study done in Kenya revealed that the most common lesions seen in patients with rheumatic heart disease were of regurgitant type with mitral valve leading, followed by, aortic valves and the least common was functional pulmonary regurgitation[11].

From the 65 patients diagnosed as having CHD, 20(35.7%) have cyanotic congenital heart disease, 31(55%) have acyanotic left to right shunt, and 5 (8.9%) with vascular malformation. This is in agreement of a study done in Nigeria upon school pupils where acyanotic CHD were found in 30 (96.8%) pupils while cyanotic CHD was seen in only one (3.2%) pupil[17].

The echo findings showed that the commonest lesion was large to moderate VSD, found in 18 patients (22.5 %). Followed by Tetralogy of fallot in 9(11%), Pulm. atresia/stenosis+/- VSD in 6 (7.5%). PDA in 4(5%), ASD and TGA with a shunt each in 3 (3.75%) as well as Single ventricle/atrium /complex cyanotic HD also represent 3.75% of the study group.

PFO, Tricuspid atresia and AV canal have the same incidence, each represent 2.5% of the study group (2 patients). Trunkus arteriosus was found in1 patient (1.5%). Similar results were obtained in previous studies in Sudan, were ventricular septal defect, atrial septal defect, tetralogy of Fallot, patent ducts arteriosus and pulmonary stenosis were the commonest diseases.[13,18] This is also similar to study that measured the pattern of congenital heart disease in 4 areas in Saudi Arabia and showed that Ventricular septal defect was the commonest disorder accounting for 33.9% of Congenital heart disease.[19] again in Nepal and Uganda VSD was the commonest.[20,21] But In the study that was conducted in Nigeria,[17] the commonest cardiac defects among the acyanotic CHD was atrial septal defects (83.9%)followed by ventricular septal defects (9.7%). 39 patients (49%) were planned for urgent or early surgery or a diagnostic and therapeutic intervention. 9(11%) needed

acute medical treatment for HF OR infective endocarditis and Long-term medical treatment and follow up was the plan in 32 patients (40%).

Anne maria et al. supporting our findings as male were at higher risk for the cardiac diseases compared with female[22]. Ravi Dhingra et al. reported age was significantly associated with cardiac diseases as our study suggests[23].

RECOMMENDATIONS

Kordofan is a wide area in Sudan and is inhabitated by a considerable population including children. Studies showed that this is one of the places worldwide with highest rates of rheumatic heart disease among pediatric age groups. Also congenital heart disease is very commonly encountered medical problem as is shown in this study. Recommendations that can be drawn from this study are very essential for government and the local ministry of health to start the constitution of a highly equipped cardiac center providing both medical and surgical services for the affected children and supplied by appropriate diagnostic tools including excellent quality diagnostic and therapeutic cardiac catheterization. This necessitates the need for the local health authority to adopt specialized and technical training of pediatricians, doctors and nursing personnel in the corresponding. Aspects of pediatric cardiology and to gather and divert the resources towards this important health issue objective. Rheumatic cardiac disease is preventable through the proper and complete antibiotic treatment of streptococcal pharyngitis and tonsillitis and it is vital to apply and strict to the recommendations of the national program of prevention of rheumatic fever and rheumatic heart disease. This should reinforce by more work on improving the socioeconomic status, housing conditions and spread of the knowledge concerning the condition among the different sectors in the community. Cooperation with different bodies caring for cardiac disease in children inside or outside the country, governmental or private and also with the nongovernmental organizations concerned with child health and direct the attention and contribution of these institutes and organizations towards share in offering solutions to the obstacles encountered in pediatric cardiac service in Krdofan and in Sudan as general.

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REFERENCES

1. Van der Linde D, Konings EE, Slager MA, et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. J Am Coll Cardiol 2011; 58(21):2241–2247.
2. Zühlke L, Mirabel M, Marijon E. Congenital heart disease and rheumatic heart disease in Africa: recent advances and current priorities. Heart. 2013; 99(21):1554–1561.
3. Marijon E, Ou P, Celermajer DS, et al. Prevalence of rheumatic heart disease detected by echocardiographic screening. N Engl J Med. 2007; 357(5):470–476.
4. Edwin F, Entsua-Mensah K, Sereboe LA, et al. Conotruncal heart defect repair in Sub-Saharan Africa: remarkable outcomes despite poor access to treatment. World J Pediatr Congenit Heart Surg. 2016;7(5):592–599.
5. Hewitson J, Brink J, Zilla P. The challenge of pediatric cardiac services in the developing world. Seminars in Thorac Cardiovas Surg 2002;14(4):340–345.
6. Dearani JA, Neirotti R, Kohnke EJ, et al. Improving pediatric cardiac surgical care in developing countries: matching resources to needs. Seminars in Thoracic and Cardiovascular Surgery: Pediatric Cardiac Surgery Annual. 2011; 13(1):35.
7. Nampijja D, Kumbakumba E, Bajunirwe F, et al. Burden of heart diseases in children attending cardiology clinic in a regional referral hospital in Uganda. Int Clin Pathol J. 2017;4(4):92-94.
8. Sulafa KM, Karani Z. Diagnosis, management and outcome of heart disease in Sudanese patients. East Afr Med J 2007; 84:434–440.
9. Ali S. Rheumatic heart disease control in Sudan: a “research for life” initiative. Sudan J Paediatr. 2018; 18:6–9. .
10. Sulafa Ali, Sara B Domi, Abdelsalam MH Elfaki, Khalid Al Talib, Mohammed H Abdelrahman, Mohamed S Adam, Mugahid MA Ahmdia. The echocardiographic prevalence of rheumatic heart disease in North Kordofan and initiation of a control program. Sudan Med J, 2017; 53(2):63-68
11. Kironget AC, Kiptoon P, Koech M. Echocardiographic Characteristics of Children with Rheumatic Heart Disease At Moi Teaching and Referral Hospital (Mtrh), Eldoret, Kenya. Eur J Biol Med Sci Res 2014; 2(4): 17-29.
12. Abah RO,Ochoga MO,Audu OP,Idoko A,Eseigbe EE,Dabit JO, Pattern of cardiac diseases among Children in a tertiary hospital in North Central, Nigeria: A three and half years retrospective cohort Echocardiographic study, Niger J Pediatric 2018; 45 (1): 6 – 9
13. El Hag A,Pattern of congenital heart disease in Sudanese children,East Afr Med J. 1994 Sep;71(9):580-6.
14. Hodes R.M, Pattern of Heart Disease in Ethiopia as Seen in a Cardiology Referral Clinic, Cardiology 1988; 75:458–464.
15. Sani UM, Ahmed H, Jiya NM. Pattern of acquired heart diseases among children seen in Sokoto, North-Western Nigeria. Nig J Clin Pract 2015; 18(6): 718-725.
16. Sani MU, Karaye KM, Borodo MM. Prevalence and pattern of rheumatic heart disease in the Nigerian savannah: an echocardiographic study. Cardiovas J Afr. 2007; 18(5): 295.
17. Ujuanbi AS, Tabansi PN, Otaigbe BE. Prevalence of Congenital Heart Diseases Among Primary School Children in the Niger Delta Region of Nigeria, West Africa. Ann Pediatr Child Health 20016; 4(5): 1116.
18. Suliman A. The state of heart disease in Sudan. Cardiovascular J Afr. 2011; 22(4):191-196.
19. Alabdulgader AAA. Congenital heart disease in Saudi Arabia: current epidemiology and Future projections. Eastern Med Health J 2006; 12 (Supplement 2).
20. Joshi A, Shrestha RPB, Shrestha PS, Dangol S,Shrestha NC, Poudyal P, Shrestha A. Pattern of CardiacDiseases in Children Attended at Dhulikhel Hospital,Nepal. Kathmandu Univ Med J 2016; 55(3):239-243.
21. Nampijja D, Kumbakumba E, Bajunirwe F, Mondo C. Burden of heart diseases in children attending cardiology clinic in a regional referral hospital in Uganda. Intern Clin Pathol J 2017; 4(4): 92-94.
22. Anne Maria Möller-Leimkühler .Gender differences in cardiovascular disease and comorbid depression. Dialogues Clin Neurosci. 2007 Mar; 9(1): 71–83.
23. Ravi Dhingra, Ramachandran S. Vasan. Age as a Cardiovascular Risk Factor. Med Clin North Am. 2012 Jan; 96(1): 87–91.