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Research Article

Clinical Outcomes of Patients Presenting in One Hospital with Congenital Heart Disease and Rheumatic Heart Disease

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Abstract

Background: Acute rheumatic fever (ARF) is a post infectious, non-suppurative sequela of pharyngeal infection with *Streptococcal pyogens*, or group A beta hemolytic streptococcus. Africa has one of the highest prevalence of heart disease in children and young adults, including congenital heart disease (CHD) and rheumatic heart disease (RHD). The incidence of CHD is about 8 per 1000 live births. The prevalence of RHD in school aged children is estimated to be in the range of 2 - 11 per 1000.

Objectives: To analyse 6 patients that presented with congenital heart disease associated with rheumatic heart disease in a hospital that is offering tertiary paediatric cardiac services.

Methods: This study details retrospective review of 6 patients from the Eastern Cape Province of South Africa that presented with congenital heart disease associated with rheumatic heart disease. We reviewed records of patients seen and/ admitted in paediatric cardiology unit at Dora Nginza Hospital with RHD and CHD from January 2008 to January 2015 (seven years). We reviewed demographic, clinical presentation, echocardiographic diagnosis of rheumatic fever/rheumatic heart disease and congenital heart disease and clinical outcomes in these patients.

Results: Six patients presented with congenital heart disease associated with rheumatic heart disease. Four were male patients and two female patients. Average age at presentation was 8.5 years. The youngest patient was 4 years old. Three patients had atrial septal defect (ASD). Two patients had patent ductus arteriosus (PDA). One patient had ventricular septal defect (VSD). The two patients with PDA's were closed per-cutaneous. And the rest of the patients were repaired surgically. They all had good results post-intervention.

Conclusion: Patients that present with rheumatic heart disease may also have congenital heart disease. Therefore, we must always lookout for congenital heart disease in patients that present with features of ARF/RHD and vice versa. The relationship between rheumatic heart disease and congenital heart disease requires further exploration with prospective studies to define its nature.

Keywords: *Rheumatic Heart Disease; Rheumatic Fever; Congenital Heart Disease; Surgery; Outcomes*

Abbreviations

ARF: Acute Rheumatic Fever; ASD: Atrial Septal Defect; CHD: Congenital Heart Disease; PDA: Patent Ductus Arteriosus; RF: Rheumatic Fever; VSD: Ventricular Septal Defect; WHF: World Health Federation

Introduction

Acute rheumatic fever (ARF) is a post infectious, non-suppurative sequela of pharyngeal Infection with *Streptococcal pyogens*, or a group A beta hemolytic streptococcus [1]. More than one third of affected children develop carditis, followed by progressive and permanent valvular lesions.

Devastating complications of rheumatic heart disease (RHD) include severe valve regurgitation, heart failure, strokes and infective endocarditis, usually affecting both younger school going and economically active, child bearing members of society [2].

It is estimated that there are over 15million cases of RHD worldwide, with 282 000 new cases and 233 000 deaths annually, which is likely a significant under-estimation according to the increasing data on subclinical RHD [3].

In Africa, RHD has demonstrated a particular prevalence in the younger African population [4].

Rheumatic heart disease present with mitral valve and aortic regurgitation. Mitral valve regurgitation is the leading indication for surgery in rheumatic heart disease. Paediatric patients presents less with mitral and or aortic stenosis [2].

The incidence of congenital heart disease (CHD) is about 8 per 1000 live births [5]. A survey conducted in Mozambique provide an opportunity to assess the prevalence of CHD in the general population of public school children in Maputo [6]. Five children (out of 2170) had CHD giving a prevalence of 2,3 in 1000 of which 80% were newly discovered.

The epidemiological patterns of heart disease differ greatly between developed nations and sub-Saharan countries, where RHD remains a public health issue in Africa while congenital heart disease bears a poor prognosis [7]. 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 [8-10].

The occurrence of congenital heart disease with rheumatic heart disease has been described [11]. In this study, the case sheets of 285 consecutive patients admitted with a diagnosis of CHD or rheumatic fever (RF)/ RHD in the one- year period beginning December 2000 were analysed. Five boys aged 8 - 12 years were diagnosed to have CHD as well as RF/RHD in this population. The diagnoses of CHD was made prior to the rheumatic affection in two patients while these disorders were simultaneously diagnosed in the remaining three patients. Thakur., *et al*. [12], have reported this phenomenon amongst school children of Shimla hills. They found that the prevalence of RF/RHD was significantly higher in children with CHD (8, 8%) as compared to those without CHD (0, 3%). Thakur., *et al*. [12], believe that the presence of CHD predisposes the child to the occurrence of RF/RHD, and special care and regular follow up of patients with CHD is required for occurrence of RH/RHD. Simultaneous diagnosis of RF/RHD with CHD will allow simultaneous correction of both the lesions during a single surgical intervention [13].

We report 6 patients that presented with a combination of acyanotic congenital heart disease and rheumatic heart disease in a clinical review of 60 patients with RF/RHD in tertiary hospital setting in South Africa.

Methods

This was a retrospective review of records of patients with RF/RHD that were seen and admitted at Cardiology Unit in Dora Nginza Hospital, Eastern Cape Province, South Africa from 2008 - 2015. Six patients (out of 60) also presented with congenital heart disease associated with RF/RHD. We analysed the 6 patients that presented with simultaneous RF/RHD and CHD separately. We reviewed clinical presentation, echocardiographic diagnosis of RF/RHD and CHD, patient management and outcomes. The diagnosis of ARF or RHD was based on clinical and echocardiographic evidence of RHD, using the newly revised Jones criteria of the World Health Federation (WHF) of 2012 [14]. In addition, an antistreptolysin O titre (ASOT) was performed in all patients as an indication for recent streptococcal infection.

Inclusion Criteria

All patients diagnosed simultaneously with RHD and CHD presenting at paediatric Cardiology, Dora Nginza Hospital between 2008 and 2015 were included.

Exclusion criteria

Patients that did not present with a combination of RHD and CHD were excluded.

Results

Out of 60 patients that were analysed for RF/RHD from 2008 to 2015, 6 patients had a simultaneous diagnosis of CHD with RHD (Table).

Features	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Age at presentation	13 years	9 years	7 years	4 years	6 years	12 years
Type of CHD	Sinus venosus ASD	PDA	PMVSD	PDA	Secundum ASD	Sinus venosus ASD
Type of RHD	Severe MR	Mild MR	Severe MR and AR	Mild MR	Severe MR	Severe MR/TR
Treatment	ASD patch repair and mitral valve repair	PDA device closure. Awaiting MV repair	VSD patch closure. MV and AV repair.	PDA device closure	ASD patch closure and MV repair	ASD closure. MV repair
Outcomes	Good results. Normal LV function post-op	Good results. Awaiting elective MV repair	Good results	Good results. Awaiting elective MV repair	Good results	Good results

Table: *Clinical profile of patients with CHD and RHD. (CHD, congenital heart disease; RHD: Rheumatic Heart Disease; ASD: Atrial Septal Defect; PDA: Patent Ductus Arteriosus; PMVSD: Peri-Membranous Ventricular Septal Defect; MR: Mitral Regurgitation; AR: Aortic Regurgitation; TR: Tricuspid Regurgitation; MV: Mitral Valve; AV: Aortic Valve.*

Four male patients and two female patients. Three patients had atrial septal defect (ASD). Two patients had patent ductus arteriosus (PDA). And only one patient had ventricular septal defect (VSD).

All the patients presented simultaneously with CHD and RHD and they were diagnosed at the same time on presentation. Majority of patients (5 out of 6) presented between the ages of 5 - 15 years, which is known to be the risk age group for RF/RHD. Only one patient presented at 4 years of age, and this was the youngest patient. The average age at presentation was 8, 5 years.

Four patients presented with severe mitral valve regurgitation, three of them had ASD, and one had VSD. Two patients with PDA presented with mild mitral valve regurgitation. No patients presented with mitral valve stenosis and or aortic valve stenosis.

Only two patient presented with severe left ventricular diastolic dysfunction (one patient had a large PDA and another had a large ASD). The rest of the patients had normal left ventricular function at presentation.

Two patients with PDA's were closed per-cutaneous with good results, both patients remained with RHD and are on medical therapy (captopril and penicillin vk) awaiting elective mitral valve surgery. Three patients had ASD and mitral valve repair with good results. One patient had VSD repair and mitral valve repair with good results. They are all doing well clinically on follow up. There was no death in this cohort.

Discussion

Rheumatic fever and rheumatic heart disease remains a burden in the developing countries, while congenital heart disease bears a poor prognosis. The Province of the Eastern Cape in South Africa is known to have high burden of infectious diseases due to high levels of poverty.15 therefore children in the Eastern Cape still suffers from RF/RHD despite decrease in the prevalence of RHD elsewhere in the country [2,16]. In our series of 60 patients with RF/RHD that were reviewed over a period of 8 years, 6 patients presented with CHD associated with RHD. All the patients met the WHF clinical and echocardiographic criteria for the diagnosis of rheumatic fever/ rheumatic heart disease [14].

This has been reported by Thakur., *et al*. [12] who reported high prevalence of RHD in patients that presented with CHD compared to patients without CHD. Thakur., *et al*. believes that the presence of CHD predisposes to RF/RHD, and that special care and regular follow up is advised for patients with CHD for occurrence of RF/RHD. In our study out of 60 patients, only 6 patients who had CHD presented with RF/RHD simultaneously. All these patients presented with heart failure symptoms and had features of rheumatic heart disease, and were also diagnosed with congenital heart disease on echocardiography.

Eighty three percent (5/6) patients presented between the ages of 5 - 15 years of age which is the high-risk age for development of RF/RHD [2]. only one patient presented earlier at 4 years of age with PDA and RHD. Bhokhandi., *et al*. [11] presented 5 patients out of 285 patients, who were diagnosed with CHD associated with RHD. All of them were males, age group between 8 - 12 years. The age group was similar to our cohort, the average age at diagnosis was 8, 5 years in our series.

All the patients presented with mitral valve regurgitation of rheumatic origin. This is the common presentation of paediatric rheumatic heart disease, and is the leading indication of rheumatic mitral valve surgery in children [17]. The fact that our patients also presented with acyanotic congenital heart disease with left to right shunting (L>R) contributed to the severity of rheumatic heart disease due to left ventricular volume overload secondary to increased pulmonary blood flow due to the L>R shunting.

Out of 6 patients only two patients presented with left ventricular diastolic dysfunction pre-operatively, one patient had a large sinus venosus ASD and severe mitral valve regurgitation and the other one had large PDA and mitral valve regurgitation. This was also due to the added effect of congenital heart disease. Patients left ventricular function improved post-operatively. Four patients presented with normal left ventricular function. Two patients who presented with PDA are still waiting for elective mitral valve surgery.

It seems patients presented with RF/RHD symptoms and signs and were also found to have CHD on diagnosis, because they almost all presented at the high-risk age for RF/RHD risk (5 - 15 years), except for 1 patient (PDA) who also presented late in terms of PDA (presented at 4 years of age). All the patients had good outcomes post-surgical (4 patients), and 2 PDA post-percutaneous closure.

Conclusion

Patients that present with rheumatic heart disease may also have congenital heart disease. Therefore, we must always lookout for congenital heart disease in patients that present with features of RF/RHD and vice versa. Simultaneous diagnosis of RF/RHD with CHD will allow simultaneous correction of both the lesions during a single surgical intervention. The relationship between rheumatic heart disease and congenital heart disease requires further exploration with prospective studies to define its nature.

Study Limitations

This was a retrospective chart audit of patients seen and admitted in one hospital.

Conflicts of Interest

None.

Permissions

Permissions granted by the CEO of the hospital.

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