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Clinical Profile of Paediatric Patients with Rheumatic Heart Disease at Moi Teaching and Referral Hospital (MTRH), Eldoret, Kenya

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Abstract
Rheumatic heart disease (RHD) is the commonest acquired heart disease and a major cause of morbidity and mortality among children in developing countries. This study sought to determine the clinical profile of paediatric patients with RHD at Moi Teaching and Referral Hospital (MTRH), Eldoret, Kenya. It employed a cross sectional study in the general peadiatric wards, paediatric outpatient clinic and cardiology clinic at the MTRH. The study subjects were children with RHD aged 3 to 15 year. In total, eighty four children (28 boys and 56 girls) with RHD were enrolled. Data was collected in a structured questionnaire and analyzed using Genstat discovery. The study found that the commonest symptoms among new patients were dyspnea, easy fatigability, palpitation, cough and orthopnea while the commonest signs were a systolic murmur, a thrill and tachycardia. Most new patients presented in NYHA class 3 and 4. Mitral regurgitation alone was the commonest lesion, followed by mitral regurgitation + aortic regurgitation. New patients had clinical evidence of severe valvular disease and complication implying late presentation. It is thus recommended that emphasis should be put on early detection and primary prevention as most new patients have severe disease.
Keywords: Rheumatic heart disease, Clinical Profile, Paediatric, Moi Teaching And Referral Hospital (MTRH)

1. Introduction

Rheumatic heart disease (RHD) is the commonest acquired cardiovascular disease in children and young adults. It accounts for 12–65% of hospital admissions related to cardiovascular disease, and for 2.0–9.9% of all hospital discharges (WHO 1992; 2001). It is a major cause of morbidity, disability and mortality in developing countries. An estimated 60 000 children and young adults die annually from rheumatic heart disease. The mortality rate per 100 000 population varied from 1.8 in the WHO Region of the Americas, to 7.6 in WHO South-East Asia Region. A case fatality rate of, 9.7% has been reported in Fiji. The annual mortality is 1.5% per year which is a conservative estimate. The most devastating effects are on children and young adults in their most productive years (WHO, 2001; Nkomo, 2007; Steer *et al.,* 1999).

Rheumatic heart disease is the most serious complication of rheumatic fever (RF), the disease which "licks the joints, but bites the heart" … to put it lightly. Rheumatic fever develops in children and adolescents following pharyngitis infection with group A beta-hemolytic Streptococcus (i.e. Streptococcus pyogenes). The organisms attach to the epithelial cells of the upper respiratory tract and produce a battery of enzymes allowing them to damage and invade human tissues. After an incubation period of 2-4 days, the invading organisms elicit an acute inflammatory response with 3-5 days of sore throat, fever, malaise, headache, and an elevated leukocyte count (WHO, 2001). After a latent period of 1-3 weeks, antibody induced immunological damage occur to heart valves, joints, subcutaneous tissue and basal ganglia of brain. The processes leading to acute rheumatic fever are still poorly understood. New data suggest that a unique surface marker on non-T lymphocytes in patients with rheumatic fever and rheumatic heart disease may prove helpful in defining which individuals are susceptible to developing rheumatic fever after a streptococcal infection because of abnormal immune responses (Steer *et al.,* 2009). Rheumatic carditis is present in 40 - 60% of patients with acute rheumatic fever; it ranges from mild to severe carditis. Cardiac involvement almost always occurs in recurrent episode. About 90% of children who have carditis during rheumatic fever episodes will develop chronic and progressive RHD from inflammation and scarring of the heart valves, which may result in haemodynamically significant valvular regurgitation and /or

stenosis, heart failure, and death (WHO, 2001).

In addition to the human suffering involved, this vicious disease can also contribute to the crippling of a country's economy. This is not only because of the medical and surgical costs of treatment, but also because it is a disease that primarily attacks children, adolescents and young adults (the most economically active group of any country) (Cilliers, 2006).

The Key to accurately understanding and diagnosing rheumatic heart disease is to get Clinical profile and real time imaging which gives more insight into the nature of the disease and which valves are affected. Currently Clinical profiles of rheumatic heart disease in children at the Moi Teaching and Referral Hospital have not been documented.

2. Materials and Methods

The study was conducted at MTRH paediatric wards, outpatient clinic and paediatric and adult cardiac/ echocardiography clinic of the Moi Teaching and Referral Hospital (MTRH) using a cross-sectional study design. Paediatric patients with rheumatic heart disease aged 3-15 years between October 2009 and October 2010 were the target population. Children aged 3-15 years were chosen because the incidence of pharyngeal infections, rheumatic fever and RHD is highest in this age group. RHD is uncommon before 3 years of age.

All children who met the clinical diagnosis and American heart association diagnostic criteria of RHD during the study period were included. A diagnosis of RHD was based on a combination of history, clinical examination, and WHO diagnostic criteria of RHD defined by an expert panel convened under the auspices of the WHO and the National Institutes of Health in September 2005. Children seen in the clinics and wards with a suspected or known diagnosis of RHD were re-evaluated by the principal investigator and those with symptoms and signs suggestive of rheumatic heart disease were selected for investigation for possible inclusion into the study. A Doppler echocardiography was performed to confirm the diagnosis using the American heart association combined criteria for diagnosis of RHD.

The author sensitized the pediatric wards, pediatric outpatient, echocardiographic laboratory and cardiology clinic staff on the study. When children with rheumatic heart disease were admitted or seen in the clinic the principal investigator was alerted and she evaluated them for possible inclusion in the study. The author then evaluated all children with a diagnosis of RHD for the following symptoms and signs; cough, haemoptysis, dyspnea, paroxysmal nocturnal dyspnea, orthopnea, chest pain, palpitation, syncope, body swelling, easy fatigability, abdominal distention, oedema, murmurs, wasting, tachypnea, cyanosis, tender hepatomegally, thrill basal crepitations, and the heart rate and rhythm. New York Heart Association Classification was used in this study to estimate the functional capacity of a patient's heart. (NYHA developed as functional classification for patients with heart disease) (The Criteria Committee of the New York Heart Association, 1994).

3. Results

3.1 Demographic characteristics

The demographic characteristics of the patients who were included in the study are presented in Table 1. A total of eighty four children with rheumatic heart disease were enrolled in the study (46 new patients and 38 on follow-up treatments).

Table 1: the demographic characteristics of patients with rheumatic heart disease

	Patient type		
	No. of patients with an initial diagnosis of RHD	No. of patients on follow up	Total
Female	32(38.10%)	24(28.57%)	56(67%)
Males	14(16.67%)	14(16.67%)	28(33%)
Total	46(54.76%)	38(45.23%)	84

Overall, there were more female patients 56(67%) than males 28 (33%), with a male to female ratio of 1:2.The median age was 11 yrs. (3.5, 15)

There were more new patients than those already on treatment and who were being followed up.

There were no significant differences in nutritional status by gender, with 50 %(14) and 52 %(29) of males and females respectively having normal weight for age. Forty three percent of both male and female were underweight while only 5% of the females and 7% of the male had marasmus.

The distribution of patients by age group and nutritional status is presented in Figure 1.

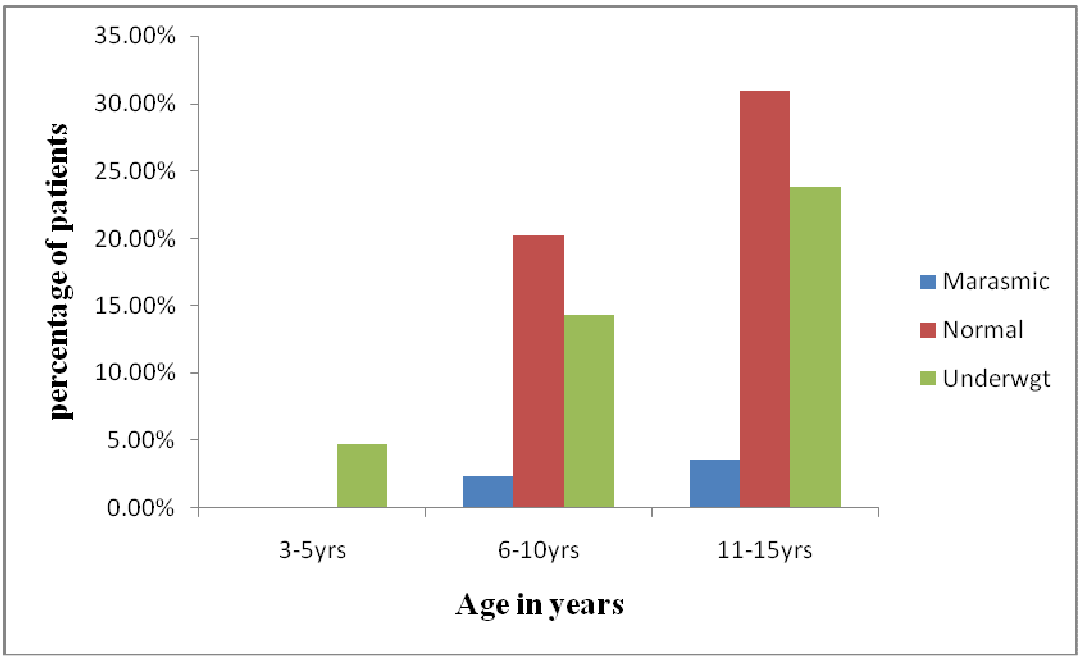

Figure 1: Percentage of patients with Rheumatic heart disease (y-axis) having different nutritional status within each age group of the population studied

There were 49(58.33%) children aged >10yrs, 31(36.90%) between 6-10yrs and four (4.76%) aged less than 5 years.

From the population studied, half of the patients (51.19%) had a normal weight for age, 42.68% were under weight and 5.95% were marasmus.

The symptoms and signs exhibited by patients with RHD for the population studied are presented in Tables 2 and 3 respectively.

Table 2: **Percentage of patients with RHD exhibiting different symptoms**

Symptoms	New patients	Patients on follow up	P value
Dyspnea	39(84.78%)	24(63.16%)	0.265[1]
Easy fatigability	38 (82.61%)	24(63.16%)	0.147[1]
Palpitation	34(73.91%)	18(47.37%)	0.089[1]
Cough	32(69.57%	14(36.84%)	0.416[1]
Orthopnea	29(63.04%)	13(34.21%)	0.660[1]
Paroxysmal nocturnal dyspnea	29(63.04%)	8(21.05%)	**<0.001[1]**
Body swelling	24(52.17%)	8(21.05%)	**0.003[1]**
Chest pain	19(41.30%)	11(28.95%)	0.175[1]
Abdominal distension	18(39.13%)	4(10.53%)	0.353[1]
Asymptomatic	6(13.04%)	8(21.05%)	0.992[1]
Syncope	6(13.04%)	0	0.014[2]
Haemoptysis	1(2.17%)	0	0.556[2]

[1]chi square test,[2] fishers exact test

The most common symptoms presented by the new patients were dyspnea, easy fatigability, palpitations, cough and orthopnea. Six (13.04%) of the new patients were asymptomatic at presentation; these patients were referrals where they were discovered to have had a murmur after presenting for other non-cardiac diseases.

Paroxysmal nocturnal dyspnea and body swelling was significantly more common among the new patients.

Table 3: Percentage of patients with RHD exhibiting different signs

Signs	New patients	Patients on follow up	P value
Systolic murmur	41(86.96%)	30(78.95%)	0.458[1]
Tachycardia	35(76.09%)	20(52.6%)	**0.025**[1]
Loud P2	29(63.04%)	16(42.11%)	0.413[1]
Edema	24(52.17%)	7(18.42%)	0.050[1]
Wasting	23(50.00%)	12(23.68%)	0.480[1]
Diastolic murmur	22(47.83%)	13(34.21%)	0.861[1]
Tender hepatomegaly	21(45.65%)	8(21.05%)	0.615[1]
Pallor	18(34.78%)	11(28.05%)	0.079[1]
Tachypnea	18(39.13%)	6(15.79%)	0.111[1]
Collapsing pulse	16(34.78%)	7(18.42%)	0.595[1]
Fever	16(34.78%)	6(15.79%)	0.913[1]
Basal crepitation	15(32.61%)	5(13.16%)	0.721[1]
Increased jugular venous pressure	12(26.09%)	2(5.26%)	0.344[2]
Irregular heart rate	2(4.35%)	0	0.285[2]
Decreased breath sound	1(2.19%)	1(2.63%)	0.437[2]
Malar flush	0	0	-
Cyanosis	0	0	-

[1]chisquare test ,[2] fisher's exact test

The most common signs exhibited by the new patients were a systolic murmur (89.13%), thrill (78.26%) and tachycardia (76.09%) of new the patients. Tachycardia was significantly more common in the newly diagnosed patients.

Numbers of patients who were in heart failure and their NYHA class are illustrated in Figure 2

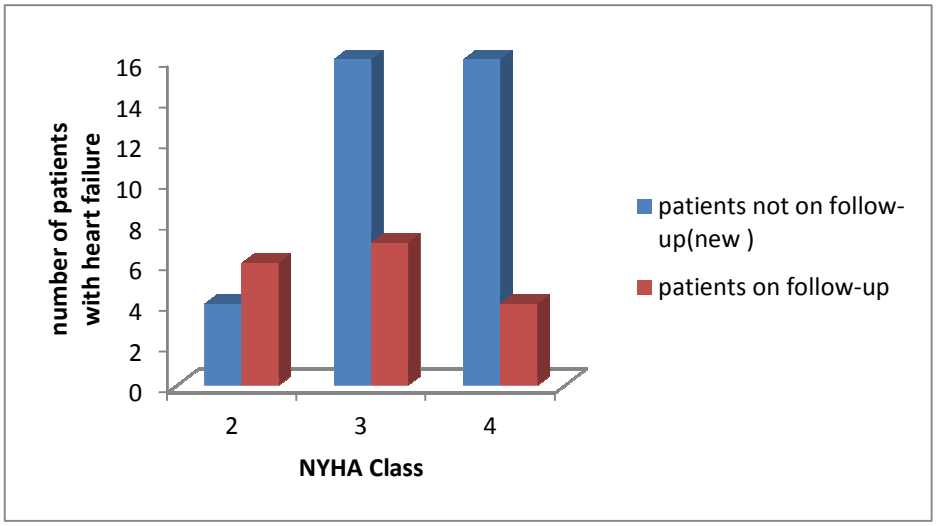

Figure 2; NYHA class of patients in heart failure in the different patient cohorts

Eighteen (47.37%) of the patients on follow-up and 36 (78.26%) of the newly diagnosed patients were in heart failure. The new patients were mainly in NYHA class 3 and 4.

3.2 New York heart association classification

A clinical classification of patients was carried out using the New York heart association (NYHA). Comparisons of the various patient characteristics within the NYHA classes are presented in Table 4.

Table 4: Proportion of patients with various characteristics within each New York heart association classification class

Characteristic	NYHA CLASS					
	1	2	3	4	Total number	P value
Gender						
Female	12(21.43%)	11(19.24%)	19(33.93%)	14(25%)	56	0.126[1]
Male	10(35.71%)	7(25%)	5(17.85%)	6(21.43%)	28	
Patient status						
New	7(15.23%)	7(15.23%)	16(34.78%)	16(34.78%)	46	<0.001[2]
On prophylaxis	15(39.47%)	11(28.95%)	8(21.05%)	4(10.52%)	38	
Total for each class	22(26%)	18(21%)	24(29%)	20(24%)	84	

[1]chisquare test ,[2] fishers exact test

Differences in the overall percentage of patients within the four NYHA classes were not large, with 26%, 21%, 29% and 24% of the patients in classes 1, 2, 3 and 4 respectively. There were significantly more new patients in NYHA classes 3 and 4, than those on secondary prophylaxis who were mainly in NYHA class 1 and 2 (p <0.001).

3.3 Compliance to secondary prophylaxis

Numbers of patients on prophylaxis and their relative compliance to the secondary prophylaxis are illustrated in Figure 3. Generally, most patients were compliant; however, patients who were on secondary prophylaxis for less than a year were more likely to be non- compliant.

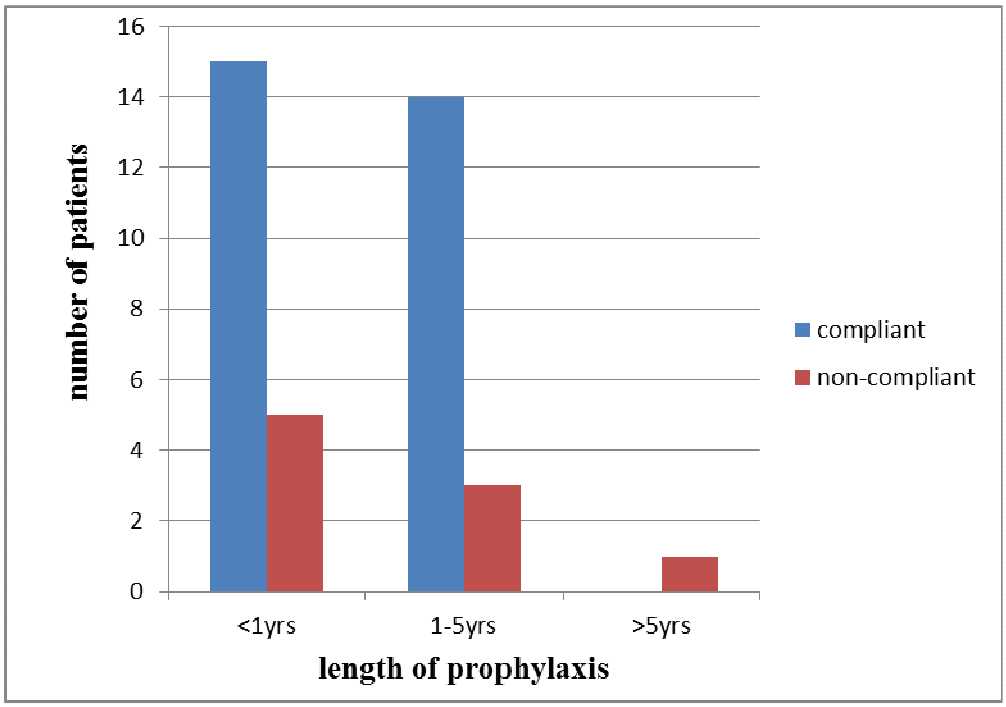

Figure 3: Duration of prophylaxis versus number of compliant and non-compliant patients

Mean duration of follow up was 14.6 month, with a range of 1 month to 6 years.

Twenty nine (76.32%) of the children were compliant while 9(23.68%) were non-compliant.

There were only 9 patients who did not adhere to secondary prophylaxis. Among these patients, the main reasons given for non-compliance were lack of finances, long distance to a health facility and lack of understanding on

the need for sustained treatment.

3.4 History of rheumatic fever

Sixty nine (86.96%) patients gave a positive history of rheumatic fever.

3.5 Correlations between main traits present in patients with RHD

The correlation matrix for the main traits present in patients with RHD is presented in Table 5.

Table 5: Correlation coefficients and probability of significance for main traits present in patients with RHD

Trait	Pulmonary Hypertension	Severity of Vulvular lesions	Left Ventricular function	Number of symptoms displayed	Number of signs displayed	NYHA class
Pulmonary Hypertension	1	0.618 (0.0000) *	0.243 (0.0261)	0.648 (0.0000) *	0.667 (0.0000) *	0.657 (0.0000) *
Severity of Vulvular lesions		1	0.145 (0.1888)	0.461 (0.0000) *	0.558 (0.0000) *	0.453 (0.0000) *
Left Ventricular function			1	0.344 (0.0013) *	0.352 (0.0010) *	0.307 (0.0052)
Number of symptoms displayed				1	0.828 (0.0000) *	0.802 (0.0000) *
Number of signs					1	0.728 (0.0000) *
NYHA class						1

In brackets are P values, * significant values

All correlations between the traits studied were positive, however, the magnitude of the correlation between the traits were different.

The correlation between pulmonary hypertension and the number of symptoms displayed, number of signs displayed and NYHA was high (p value, 0.0000), indicating that the greater the number of signs and symptoms displayed by patients, the greater the likelihood of the patient having pulmonary hypertension. However, larger studies need to be done to find out if these correlations are significant.

4. Discussions

4.1 Demographic characteristics of patients with rheumatic heart disease

Rheumatic heart disease continues to be a major health problem in the developing countries. It accounts for a large percentage of cardiovascular disease related paediatrics admissions in Kenya. The last prevalence study that was done in 1992-1994 in Kenya showed a high prevalence (27/1000) of RHD, results which are consistent with recent studies in other developing countries (Steer *et al.,* 1999). However; this is in contrast to its virtual extinction in the developed world.

Results from this study revealed that there were more females, than male with RHD, with a male to female ratio of 1:2. This is similar to Karen Sliwa's *et al,.*2010 study in Soweto, South Africa where African females had a prevalence of 68%, Essein *et al.,* 2008. in Nigeria and Paar *et al.,* 2010 in Nicaragua. Other recent studies (Halim, & Jacques, 1961; Sadiq *et al.,* 2009; Paar *et al.,* 2010; Aly *et al.,* 1997; Danbauchi *et al.,* 2004; Folger *et al.,* 1992; Sani *et al.,* 2007; Ayub, 2009; Veasy *et al.,* 1994; Siddig *et al.,* 1992 and Essien, 2008) have however depicted either no difference in the prevalence of RHD amongst males and females, or depicted females as the dominant patient population while others depicting males as the most predominant gender. Rheumatic fever and RHD are more severe and have a worse prognosis in females than in males. Women produce more vigorous immune response and increased antibody production and estrogen significantly increases proinflammatory cytokine productions thus the elevated immune response in females may even further amplify the adjuvant effect of infection thus putting them more at risk of chronic autoimmune diseases (DeLisaFairweather & Rose, 2004). This may account for the high number of females in this study, because they are more likely to be symptomatic and hence brought into hospital for treatment (Chin *et al.,* 2010).

Children aged less than 5 years were the least prevalent forming 4.8% of those with RHD while those aged 5-

15yrs had a prevalence of approximately 95%. This is similar to what was found by Yuko- Jowi at Kenyatta National Hospital who found that 76.4% of cases were aged between 5 and 15 years, while only 3% were less than 5 years old (Yuko-Jowi, 2005).Almost half of children studied had a weight that was less than 80% weight for their age (either underweight or marasmic). This could be because of the disease process. Patients with rheumatic heart disease, especially mitral valve disease, are most likely to have cardiac cachexia, but nearly all patients with cardiac disease demonstrate wasting of body fat and skeletal muscle, with muscular weakness and easy fatigability. This is due to disturbances in neuroendocrine, inflammatory, and metabolic systems; anorexia from chronic illness, the unpalatability of a low salt diet, and to a great extent to poor cardiac output due to heart failure. In this study, evidence for cardiac cachexia may be seen in the fact almost half of the patients had advanced tricuspid regurgitation which is one of the major criteria for diagnosis of cardiac cachexia in patients with mitral valve diseases. Sadif in Pakistan found a higher prevalence (67%) undernourished children with RHD (Sadiq *et al.,* 2009).

4.2 Clinical profile of patients

The commonest symptoms were dyspnea, palpitation and easy fatigability, while the commonest signs were systolic murmur, tachycardia and a thrill. Thirteen percent of the new patients were asymptomatic they came into MTRH as referrals where they were discovered to have had a murmur after presenting for other non-cardiac diseases/symptoms.

Almost three quarters of the patients were symptomatic. This is contrary to what would be expected in a population where the commonest valvular lesion is isolated MR. In most cardiology literature patients with mild to moderate MR are usually asymptomatic. The reason for this difference is that most patients in this study had severe valvular disease with complications of pulmonary hypertension and also because MTRH being a referral hospital receives very sick patients. Daniel Bernstein reported that easy fatigability and dyspnea is present in over 70% of the patients, this corroborates with what was found in this study. Fatigability and dyspnea are common in patients with severe MR and moderate to severe MS (WHO, 2001).

There was an almost equal distribution of patient in all the NYHA classes when all the patients were analyzed together. Almost three quarters of the new patients came in stage 3 and 4. . There was an association between the NYHA class and severity of valvular lesion. Only 13% of the new patients were asymptomatic showing that most patients seek medical attentions when they are very sick and probably having severe disease.

The mean duration of secondary prophylaxis was 14.6 month and a range of 1 month to 6 yrs. Kimberly-Leaky in Brazzaville found compliance of 75% after one month of follow-up and 37.5% after 3 month of follow-up, this is contrary to what was found in this study where patients who were on prophylaxis for a longer duration were more compliant than patients who were on it for a shorter duration (Kimbally-leaky, 2008). Almost a quarter of the patients on prophylaxis were non-compliant with lack of knowledge, lack of finances and long distance to nearest health facility being the main reasons for non-compliance. There were however few non-compliant patients (9 patients). This study was a cross-sectional study hence could not find out the dropout rates from secondary prophylaxis which is an important parameter (Alkhalifa *et al.,* 2008).

4.3 Correlations between main traits present in patients with RHD

The greater the number of signs and symptoms displayed by patients, the greater the likelihood of the patient having pulmonary hypertension. There was also a positive and strong correlation between the New York heart association and the severity of pulmonary hypertension. This is consistent with the WHO classification of pulmonary hypertension which is a modification of New York heart associations where patients who are more symptomatic and have right sided heart failure have a worse WHO pulmonary hypertension classification (WHO, 1998).

5. Conclusion and Recommendations

From the study findings, the commonest symptoms of new patients at MTRH were dyspnea, easy fatigability, palpitations, cough and orthopnea while the commonest signs were systolic murmur, thrill and tachycardia. A significant number of patients were asymptomatic. Patients who were non-compliant to secondary prophylaxis were more likely to have severe valvular lesion and pulmonary hypertension.

From the study findings most of the new patients had severe disease at diagnosis; therefore, it is recommended that emphasis should be put on early detection and primary prevention. This entails eradication of pharyngeal streptococci through appropriate antibiotics therapy and long-term secondary prophylaxis to guard against recurrent pharyngeal streptococcal infection for children with acute rheumatic fever.

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