Patients Referred to the Paediatric Cardiology Outpatient Services at Red Cross War Memorial Children's Hospital in 2018:

A review of demographics, referral patterns, diagnoses and clinical outcomes

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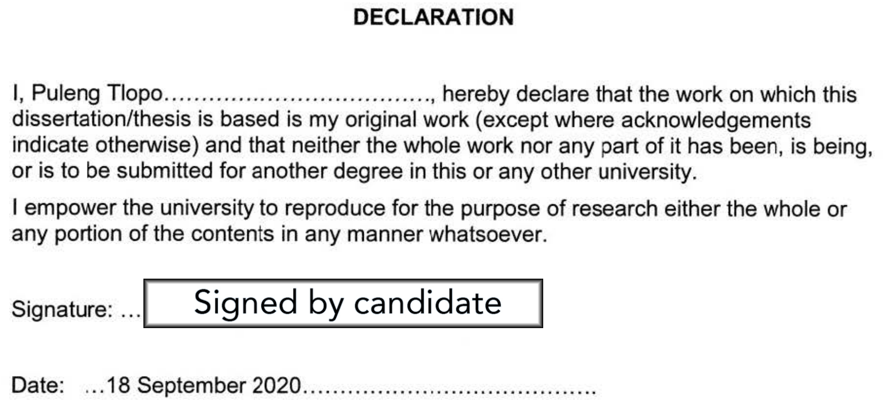

DECLARATION

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Abstract:

Background: Cardiovascular disease (CVD) in children causes considerable morbidity and mortality in Africa and its impact on health systems is an emerging problem. Paediatric cardiology is a highly specialized field based in tertiary centres with worldwide demand exceeding available resources. Upon suspicion of CVD, specialist investigation, early intervention and follow-up are major challenges in low- and middle-income settings.

Aim: To investigate and describe patients referred for specialist cardiology services at a tertiary children's hospital in Africa

Methods: Red Cross War Memorial Children's Hospital (RCWMCH) is a tertiary paediatric hospital in Cape Town, South Africa. We reviewed patients referred to our outpatient cardiology service, investigating demographics, spectrum of disease, and clinical outcomes.

Results: Between 1 January and 31 December 2018, 625 new patients were referred to cardiology outpatient department (COPD) services. Eighty percent (n=501) had structurally normal hearts. The majority of the remainder with confirmed CVD (98%, n=122) had congenital heart defects. Median age of the cohort with CVD was 3.9 months (IQR: 2.6-12.0) with a slight female preponderance (55%). Thirty-four (28%) patients were scheduled for surgery. Ten percent of referred patients had incomplete letters of referral, furthermore we found no feedback plans to these referring practitioners from the COPD doctors.

Conclusion: Demands on paediatric cardiology services at RCWMCH are high. Most referred outpatients have structurally normal hearts, straining an already overwhelmed specialist service. However, 28% of patients found to have congenital defects required surgery, highlighting the need to prioritise certain patients. Strengthening communication and feedback channels between cardiologists and generalists will optimise care.

Keywords: Paediatric cardiology referrals; Low- and middle-income paediatric cardiology services

Introduction

Cardiovascular disease (CVD) in children is a critical public health concern in Africa with congenital heart disease (CHD) and rheumatic heart disease (RHD) being the most prevalent (1). RHD remains one of the leading cause of cardiac failure and premature deaths worldwide (2). CHD is a leading cause of death in the first year of life and patients with critical cardiovascular disease will die without timeous intervention (3). It is estimated that 85% of children with CHD can reach adulthood with proper cardiovascular care while inadequate care poses a burden on patients and their families, and has deleterious economic implications (3) to both the patient's family and the the public health care system.

Paediatric cardiology services are largely based in tertiary hospitals. Referrals to these centres appear to be increasing worldwide with demand exceeding available resources (4). In 2010 Hoosen et. al. (5) described 24 paediatric cardiologists practising in a population that required at least 88 to meet international requirements. The audit reports that 1370 patients with CHD were operated on between the public and private sectors in the same year, fewer than half of the 4000 surgeries needed based on epidemiological estimates. The study suggested that less than 40% of children with CHD are receiving appropriate care with the main contributing factors being missed diagnoses, an overwhelmed paediatric cardiology service to assess referred patients timeously, and insufficient surgical and post-operative care services (5).

Upon suspicion of a cardiac lesion, the ideal management of patients is early referral, evaluation, possible intervention and follow-up by a paediatric cardiologist (6). Early recognition and intervention alter the natural progression of disease, leading to more favourable outcomes. Because of the complexity of CVD in children, there is often a need for specialist care and follow-up even after good surgical and medical outcomes (3).

As reported in the 2019 University of Cape Town Children's Institute *Child Gauge* report (7), children in sub-Saharan Africa face higher risks of dying before their fifth birthday from diarrhoeal disease, pneumonia, HIV/AIDS, neonatal conditions and injuries at a rate of 32 per 1000 live births (7). Congenital anomalies constitute 3% of these deaths with CHD being the commonest birth defect worldwide.

In the quest for optimal cardiac care for patients with CVD, patient characteristics, referral patterns and intervention outcomes are important. These factors differ between high-income countries and both low- and middle-income countries. In high-income countries, the median age of referral is lower, access to cardiothoracic surgery is easier with an increasing trend of new referrals for paediatric cardiology attention every year. A study conducted at a tertiary outpatient department in Leeds, UK, in 2005 reports 526 new referrals over a three-month period with 372 (71%) patients confirmed to have structurally normal hearts (4). Another similar study from Birmingham Children's Hospital, comparing referrals in 2006 and 2013 showed a 20% increase in referrals with most of the referred patients having insignificant pathology. This has significant implications on available resources (8).

In comparison, a study conducted at the Kenyatta National Hospital in Nairobi, Kenya, demonstrated that a large proportion of referred patients had significant pathology with most at risk for irreversible pulmonary vascular changes and therefore not amenable to surgery. Cardiac diagnoses were suspected early by generalists but confirmed late due to geographical distances and overwhelmed cardiology services in the public sector (6). Similar findings were reported from Malawi on patients referred to their out-patient cardiac service. Patients with CHD and RHD presented late with established pulmonary hypertension mostly attributed to misdiagnoses, geographical limitations and inadequate resources (9).

Given these challenges in achieving optimal paediatric cardiology care in low- and middle-income countries, understanding disease patterns, referral pathways and care gaps is essential to improve services for children with CVD and identify potential interventions.

Aims and objectives

The aim of this study was to conduct a retrospective review of all paediatric patients who were referred to the Cardiology Outpatient Department (COPD) at the Red Cross War Memorial Children's Hospital for cardiac diagnostic workup over a one-year period. The objectives were to: 1) investigate the spectrum of disease of all new patients referred to the COPD, 2) describe the age at referral and sources of referral, 3) describe the final cardiology diagnosis after full cardiological assessment and ICD-10 coding, 4) review the cardiology and management outcomes such as cardiac catherisation, surgery, electrophysiological procedures, further investigations and interventions, or referral to palliative care and finally, 5) describe mortality of new patients.

Methods:

Setting: Red Cross War Memorial Children's Hospital (RCWMCH) is a tertiary paediatric hospital in the Western Cape Province of South Africa with a fully functional paediatric cardiology service. The COPD receives between 400-600 referrals every year (excluding the critically ill patients who were not included in this study). All new patients referred to COPD are seen by a cardiology consultant, senior registrar, or cardiology registrar and routinely have an electrocardiogram, chest X-ray and echocardiogram prior to clinical review.

Data collection

Patient details were captured on a Clinicom Patient Management System, the appointments database for patients at RCWMCH. The Cardiology Department Management and Repository database provided details of echocardiographic findings, cardiac catheterisation procedures, and surgical interventions. For any further information, COPD records, called "cardiac buffs" and the hospital patient records were also reviewed. Findings were recorded on a hard copy case report form and then entered into a REDCap, secure online database hosted by the University of Cape Town.

Inclusion criteria

Our study included paediatric patients aged from birth to less than 14 years of age, referred to COPD as "new" patients (defined as patients seen for the first time by a paediatric cardiologist at RCWMCH) between 1 January 2018 and 31 December 2018 and diagnosed with congenital or acquired heart disease (confirmed by echocardiogram and including arrhythmias).

Exclusion criteria

Patients 14 years of age and older or with normal hearts

Data Analysis

Data was analysed using STATA 15.0®(10) Continuous variables are expressed as means with standard deviations or medians with interquartile ranges as appropriate. Categorical variables are expressed as frequencies and percentages.

Ethics

Permission to conduct the study was obtained from the University of Cape Town's Human Research Ethics Committee (UCT HREC 206/209; PI: HP De Decker). A waiver of consent was granted for this study as the patient population was drawn from an existing registry of paediatric cardiology patients at RCWMCH. Patients/parents seen by the paediatric cardiology service provide informed consent for demographic and medical information to be entered and stored in the registry database.

Results

Between 1 January and 31 December 2018, 1089 patients were referred to COPD. A total of 124 patients (11.4%) met our inclusion criteria.

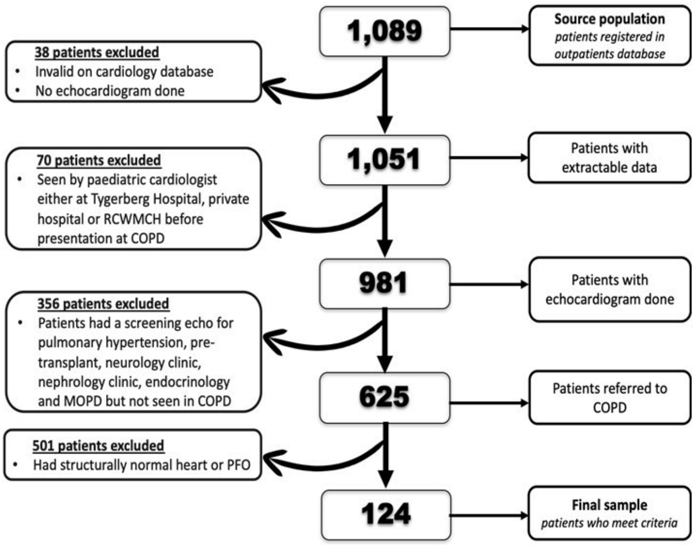

Figure 1: Recruitment of the study sample

The sample was comprised of paediatric patients, median age of 3.9 months (IQR: 2.6-12.0) with a female preponderance 55% (68/124), male 45% (56/124). The majority of referrals were from the Cape Town Metropole 98% (121/124), two patients were from other Western Cape Province localities, and the remaining patient was from the Eastern Cape Province.

Table l: Sources of referrals for patients diagnosed with congenital heart disease

Source	**n**	**%**
Level 2 hospitals	36	29.03
Other*	29	23.39
Neonatal Units	19	15.32
Community Clinics	14	11.29
Not documented	12	9.68
General practitioners	5	4.03
Private paediatricians	4	3.23
Level 3 Hospital•	4	3.23
Medical Emergency at RCWMCH	1	0.81

*Other: Genetics division, Endocrinology, Neurology, Pulmonology, ENT department, paediatric surgery

• Level 3 Hospital: Tygerberg Hospital-not seen by a cardiologist

Patients were referred by means of a referral letter. The majority of patients (83%) were referred from state-funded facilities within our catchment area such as Level 2 hospitals (Victoria and New Somerset Hospital), other sub-specialist divisions within RCWMCH, neonatology services, community clinics and those not seen by a cardiologist at Tygerberg Hospital. Seven per cent were from the private sector (4% from general practitioners, 3% general private paediatricians); 10% of patients had no documentation of the source of referral.

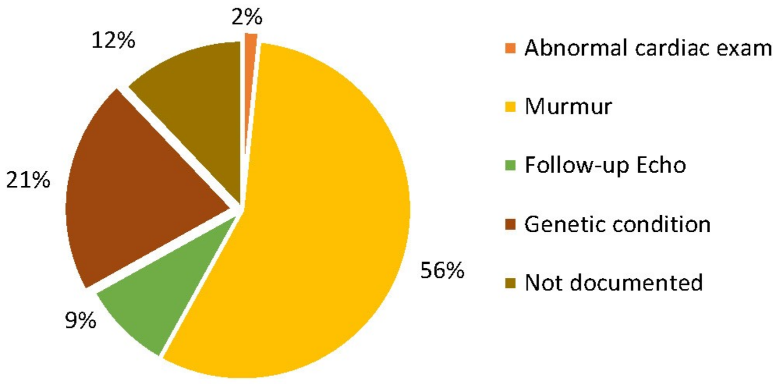

Figure 2: Reasons for referral (%)

The most common reason for referral was heart murmur in 56% of cases.

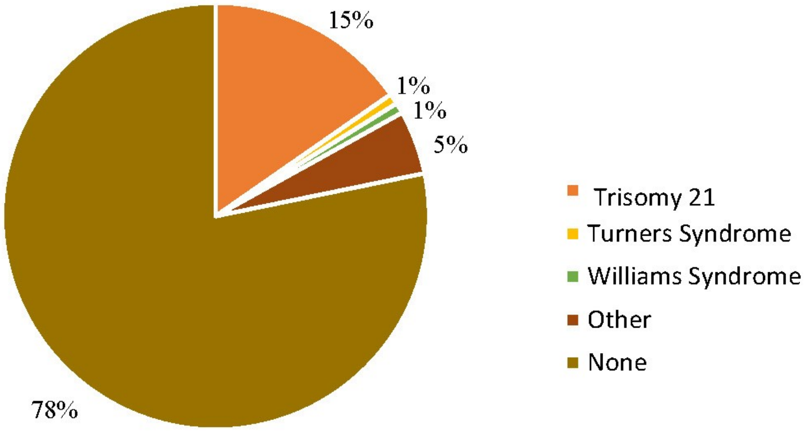

Figure 3: Associated syndromes/genetic conditions (%)

Almost a quarter of the children (21%) had associated genetic anomalies and of those, Trisomy 21 was the most common.

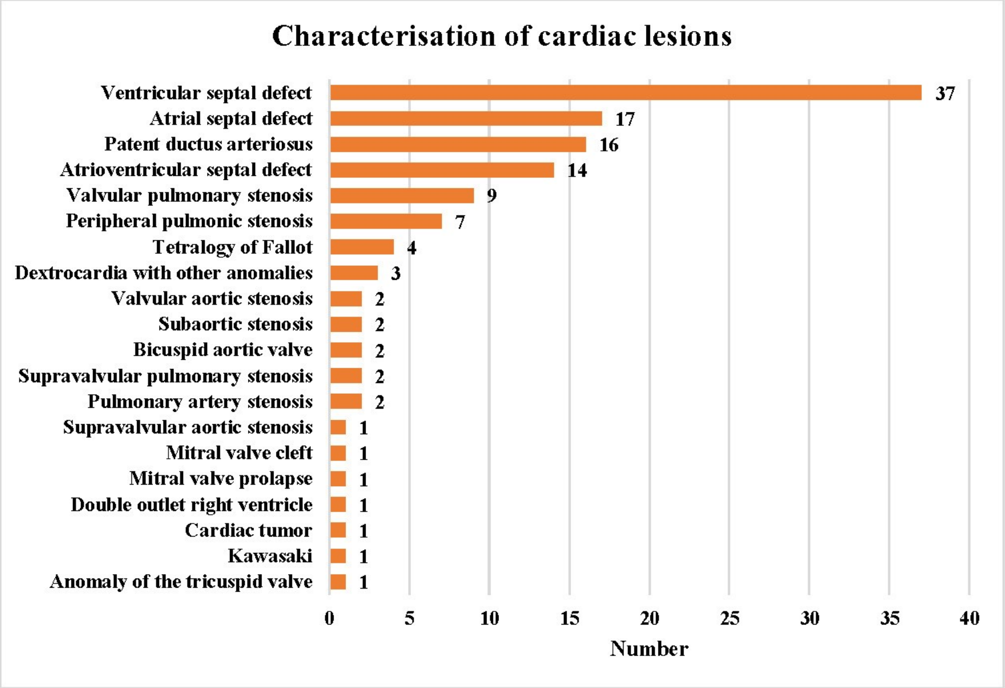

Figure 4: Characterisation of Cardiac Lesions

The majority of patients, 98% (122/124), had CHD mostly presenting with left to right shunts, with one patient having an associated Long QT syndrome. We found no patients with RHD or cardiomyopathies. One patient had a cardiac mass and one had Kawasaki disease.

Subsequent to their appointment, 28% (34) patients of those with CHD were placed on the surgical list for a procedure. The median waiting time was 104 days (Interquartile range: 43193). Of these, 97% (33/34) underwent full repair and one patient (3%) had a palliative Glenn shunt for double outlet right ventricle. The same patient had a cardiac catheterisation procedure post Glenn shunt. There were no other patients that required cardiac catheterisation.

The remaining patients were given follow-up appointments for review and 8% were subsequently discharged. Three patients were documented to have died within a year of their cardiology appointments: one patient had severe pulmonary hypertension, one had an hypoxic event unrelated to the cardiac disease and the cause of death of the third was not documented.

Discussion:

The major findings of this study are as follows: Of the patients referred to RCWMCH COPD, 80% had structurally normal hearts with an innocent cardiac murmur while 20% had confirmed CVD. CHD is the most commonly occurring cardiac lesion seen in the COPD with the majority of patients presenting with left-to-right shunts and requiring surgery within the first year after referral. Trisomy 21 is the commonest occurring genetic disorder in COPD. There are notable gaps in the referral of patients with CVD: in particular, incomplete referral letters to the clinic and no robust feedback system to referring practitioners.

There were 625 new patients referred to our COPD for evaluation by a paediatric cardiologist over the one-year review period; 56% of these patients were referred because of a cardiac murmur. Approximately 80% (501) of the patients were found to have structurally normal hearts and the remaining 20% (124) had confirmed cardiac disease.

These findings are comparable to those from high-income countries where referral of patients with structurally normal hearts for evaluation is an increasing trend. In a study done in Leeds, 71% of patients referred were reported to have innocent murmurs (4) and 60% in another study done in Ireland with a subsequent increase to 83.7% in a repeat study 5 years later (11). The main concern is that generalists are reluctant to take care of patients with features that may suggest a CHD without an echocardiogram. Interestingly, this is slightly different than findings from low-income countries where most of the patients referred have established disease and complications (6, 9).

Our study was conducted in a single centre, which may differ from other cardiology centres in South Africa. The referral of patients with structurally normal hearts to tertiary centres is reportedly increasing and has implications on patient load and the allocation of resources. As previously mentioned, South Africa does not meet its demands for cardiac care and overwhelmed cardiac services is a contributing factor.

Some recommendations in high-income countries have been the utilisation of paediatricians who have interests in cardiology to do echocardiograms before referral to tertiary services. In our setting, the majority of referrals were from level 2 hospitals run by paediatricians and less from areas outside Cape Town Metropole where there is a strong outreach programme run by RCWMCH Cardiologists. Incorporation of level 2 hospitals in the existing outreach program with scheduled clinics by paediatric cardiologists might help reduce unnecessary referrals and also serve as an education platform for paediatricians and general practitioners in those areas. Unfortunately, this is not possible due to current resource constraints.

The commonest occurring cardiac lesion in our study was CHD with the majority of patients presenting with left-to-right shunts, the commonest being a ventricular septal defect. We had no patients with RHD or cardiomyopathies as they usually present as emergencies in heart failure and are managed as inpatients. However, we did have one patient with a cardiac mass and another with Kawasaki disease which are relatively rare in our setting.

Looking at outcomes, 28% of patients with CHD required surgery and were placed on the surgical list. Two thirds of those requiring surgery had a primary repair within the first 6-12 months of referral and one patient had a palliative procedure. Interestingly, 15% (19) of the referred patients had Trisomy 21 and were therefore at risk for pulmonary vascular disease early on necessitating early surgical intervention and requiring referral within the first 6 weeks of life.

The implications of this are high demand on cardiothoracic interventions and intensive care which are very scarce resources in middle and low-income countries (5). Patients with left-to right shunts, especially those with associated genetic disorders, e.g. Trisomy 21, are at high risk for pulmonary vascular disease and need prioritisation and raised awareness amongst primary health care practitioners with active programs and policies to help optimise care.

Most patients (85-90%) in South Africa are treated in government-funded state hospitals and the remaining 10-15% in private hospitals. The COPD at RCWMCH receives approximately 400-600 new outpatient referrals every year by means of a referral letter. In our study 83% were referred from state-funded facilities including other departments within RCWMCH and 7% from the private sector (4% from general practitioners, 3% general private paediatricians). The remaining 10% had no documented source of referral. Because of differences in health systems, it is difficult to compare referral sources and the challenges they experience in referring patients for sub-specialist services at tertiary hospitals; perhaps surveys can be conducted to better understand their experiences and how these can be improved.

Patients are referred by means of a referral letter with little or no prior discussions with cardiologists. Based on our analysis, the information is often insufficient and lacking comprehensive assessment of patients. There seems to be no communication between cardiologists and primary health care practitioners. In other centres, the use of electronic consults has been demonstrated to increase primary health care practitioner access and reduce specialist waiting times for nonurgent referrals (12).

There also seems to be good documentation of patient information with specific questions to the relevant specialists thereby improving efficiency of care (12). While we do not yet utilise electronic consults, communication needs to be improved. Referral letter templates, Vula medical referral application (WWW.Vulamobile.com) and the use of the South African Department of Health *Road to Health* booklet as a communication tool to improve service and optimise care for children with CVD.

Strengths and limitations

This was the first attempt at characterising new referrals to COPD in our setting. The study highlights some of the gaps in our referral systems which are: referral of patients with insignificant pathology, poor documentation on referral letters and a limited feedback system to referring practitioners. It reveals the needs for continued education for primary health practitioners as CVD in children is very complex. It also suggests the incorporation of the referring Level 2 hospitals in the existing outreach programme which could potentially reduce the number of referrals to RCWMCH COPD.

However, a retrospective review is reliant on medical records and therefore has the inherent limitation of missing data. Referral letters were usually incomplete and it would have been useful to include a questionnaire or survey of referring physicians to get more insight into their experience of referring patients to a tertiary centre. The study was only performed over one year making it difficult to have a comprehensive review of outcomes.

Implications for practice, policy and future research

In conducting this study, we have realised the need for continued medical education from cardiologists to educate surrounding physicians on proper assessment of cardiac murmurs in tertiary hospitals, Level 2 hospitals and by general practitioners. We have also realised the need for emphasis on early referral of patients at risk for pulmonary vascular disease especially those with co-morbidities. Finally, we emphasize the need to improve the efficiency of referral systems by using referral electronic media and other resources available as communication tools between sub-specialist services and other centres.

The study allowed us the opportunity to review patient records in COPD and to suggest proper record keeping and capturing of patients on the cardiology database with clear intervention plans. Research on the utilisation of screening clinics run by cardiologists or the use of telemedicine when referring patients to tertiary services may be helpful in an effort to reduce the number of unnecessary referrals.

Conclusions

The demands on paediatric cardiology services at RCWMCH are high. The majority of patients referred to COPD have structurally normal hearts and this places a strain on an already overwhelmed specialist service. However, 28% of those with CHD required surgery within the first 6 - 12 months of referral. These are the children we should be prioritising with active programs to ensure they are referred promptly. Strengthening communication and feedback channels between cardiologists and surrounding primary health care services can help improve the service.

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Author Contributions:

Dr Tlopo designed the study protocol. She collected and entered all study data, interpreted the data analyses, compiled the data and is the primary author of the manuscript.

Prof R De Decker conceptualised the study, provided feedback on the development of the protocol and assisted with ensuring that all cases were captured and recorded during data collection. He also assisted in reviewing the protocol and manuscript.

Prof Zühlke conceptualised and designed the study, coordinated and supervised data collection, conducted statistical analyses, reviewed and revised the manuscript.

Ms Perkins assisted with protocol development, creating the data collection forms and obtaining HREC approval. Throughout the study, she assisted with logistics for collecting and entering data, and then proofread and edited the manuscript prior to submission.

Mr Basera constructed and managed the REDCap™ database, extracted the data and conducted statistical analyses. He contributed toward proofreading and revising the statistical results.

All authors reviewed and agreed upon the final submitted manuscript.

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