Original Article

INCIDENCE, TYPES, MANAGEMENT AND OUTCOMES OF CONGENITAL HEART DISEASE IN THE PAEDIATRIC POPULATION AT COLONIAL WAR MEMORIAL HOSPITAL, FIJI – FROM JANUARY 2012 TO DECEMBER 2016

Maryanne Kora'ai[1] , Kannan Sridharan[2] , Ranu Anjali[3] , Joseph Kado[4]

1. Paediatric Registrar, Paediatrics Department, Colonial War Memorial Hospital (CWMH), Fiji.
   Correspondence email: koraaimaryanne@gmail.com
2. Associate Professor, Department of Pharmacology and Pharmacy, Fiji National University, Fiji.
3. Principal Medical Officer, Paediatrics Department, CWMH, Fiji.
4. Associate Professor and Paediatric Consultant, Fiji National University, CWMH, Fiji.

ABSTRACT

Introduction:
Congenital Heart Disease (CHD) refers to structural or functional heart disease present at birth. CHD continues to cause morbidity and mortality in Fiji. There is no published data on CHD in the Pacific in the last 15 years. Unpublished data by Mataika. R, 2012 (12) estimated the incidence of CHD at Colonial War Memorial Hospital (CWMH) was ~ 0.5% or 4.9/1000 live births. This study aims to collect and document data on the incidence, types, management and outcomes of CHD, a platform upon which decisions for policy and practice development for CHD screening and management in Fiji can be made.

Method: This is a five-year descriptive, retrospective, single-centre study at CWMH.

Results:
The incidence of CHD was 5.8 per 1,000 with an uncorrected incidence of 7.2 per 1,000. Ventricular Septal defects (VSD) and Tetralogy of Fallot (TOF) were the most common acyanotic and cyanotic lesions respectively. The total number of cases of CHDs was 202 and 76 (39%) died within that period. Ninety-five percent of the deaths occurred within the first year of life. Sixty-five percent of these infant mortalities were known acyanotic CHD cases. Forty-six (23%) cases had corrective surgeries. Survival rate for surgery locally was 98%, compared to 100% for those cases that had surgery abroad.

Conclusion:
This study highlights the mortality associated with congenital heart diseases in Fiji and the need for strengthening of surgical management strategies.

Key Words: *Congenital Heart Disease, incidence, outcomes, mortality*

INTRODUCTION

Background Knowledge

Congenital Heart Disease (CHD) refers to structural or functional heart disease present at birth [1]. The aetiology may be multi-factorial; due to a single gene or chromosomal gene mutation, exposure to teratogen, environmental factors or idiopathic [2]. CHD occurs in nearly 1% of live births and is estimated to be the cause of 10% of spontaneous abortions [1]. In developed countries, foetal anomaly scan including a screening heart ulaatrasound is routinely done at 18 weeks of gestation [3] and if CHD is identified, the parents are counselled, and

corrective surgery offered in-utero or soon after birth if needed.

Screening for critical CHD continues in the early postnatal period with the use the pulse oximetry. Critical CHDs depend on the patency of the ductus arteriosus and urgent surgery during neonatal period [4]. The critical CHDs are Hypoplastic Left Heart Syndrome (HLHS), Pulmonary Atresia (with intact septum), TOF, Total Anomalous Pulmonary Venous Return (TAPVR), Transposition of the Great Arteries (TGA), Tricuspid Atresia (TA) and Truncus Arteriosus [4]. The critical CHD may go unnoticed clinically if not oximetry is not used [5].

Transthoracic and transoesophageal echocardiography are the gold standard for diagnosing CHD in children but require highly skilled personnel. Despite advances made in the diagnosis and treatment of CHD, it remains the leading non-infectious cause of death in infancy in the developing countries like Fiji.

In Fiji, with a population of about 900,000 and annual birth cohort of about 20,000, it is estimated that there are about 200 children born with CHD each year, however, there is no foetal screening or routine postnatal screening done for critical CHD. Children are referred for an echocardiogram if CHD is suspected clinically. Symptoms include cyanosis, persistent tachycardia or tachypnoea, murmurs, or if they appear syndromic.

Fiji consists of 322 islands. Eighty-seven percent of the population live on the two largest islands of Viti Levu and Vanua Levu. The main ethnic groups are i-Taukei and Fijians of Indian descent (FoI). [5]. The public health service in Fiji is divided into 4 divisions, with three divisional hospitals, CWMH in Suva, Lautoka hospital (Viti Levu) and Labasa hospital. (Vanua Levu) There are 17 sub-divisional hospitals, 80 health centres and 96 nursing stations situated across the nation. This is supplemented by three private hospitals and an increasing number of privately owned clinics operated by general practitioners.

Referrals for echocardiograms for suspected CHD are made from all levels of the health service, the school health team and from general practitioners.

Transthoracic echocardiography is the standard diagnostic investigation for confirming CHD at CWMH. Fiji has only one overseas-trained person. There are few paediatric registrars who are able to conduct screening echocardiograms on children with suspected CHD.

Children with high risk and critical CHD are referred to overseas paediatric cardiothoracic centres depending on availability of funding, while children with non-urgent, low risk cardiac lesions await the fly-in-fly-out (FIFO) cardiac teams and surgery performed locally.

Currently, three FIFO cardiac-surgical teams visit Fiji annually to perform specialist consultations and offer cardiac surgery to children with CHD. These include Open Heart International (Australia), Hearts4Kids / Friends of Fiji (New Zealand), and the Sri Sathya Sai Sanjeevani Hospital (India) team. Each team performs about twenty open heart surgeries per visit with the support from MOH, Adventist Health, Fiji Children's Heart Foundation, Sai Prema Foundation, Fiji, Corona Worldwide, Fiji Branch and other NGOs.

STATEMENT OF THE PROBLEM

CHD continues to contribute to morbidity and mortality and causes economic burden in Fiji. Children die or are admitted repeatedly for complications associated with CHD. There is lack of data on the incidence, types, management and outcome of CHD in the children at CWMH.

There is a need for data on CHD to assist in planning for the future. This study aims to collect data on the

incidence; types, management and outcomes of CHD thus creating a platform on which decisions can be made regarding the way forward for screening and management of children with CHD in Fiji

AIMS & OBJECTIVES

Aim:

To identify the incidence, types, management and outcomes of CHD in the Paediatric population at CWMH from 1st of January 2012 to 31st of December 2016.

Objectives:

- To determine the incidence of CHD in children at the CWMH.
- To identify the most common types of CHD in the children at CWMH.
- To determine the basic medical (medications) and surgical management of the children at CWMH.
- To determine the outcome (associated complications, prognosis 6 months post-surgery and further prognosticate) of children with CHD at CWMH.
- To establish and document the baseline data of CHD in the children at CWMH.

REVIEW OF LITERATURE

CHD is the most common congenital abnormality found in humans. These cardiovascular anomalies are common birth defects and a leading cause of infant mortality, morbidity and economic burden [7].

The reported incidence of CHD is about 1% according to reports from several studies from different parts of the world [2,5,6,7,8]. It is believed that this incidence has remained constant, worldwide. Nearly one third to a half of these CHD cases are critical, requiring intervention in the first year of life. Almost one third of congenital heart abnormalities are ventricular septal defects (VSDs), but atrial septal defects (ASDs), pulmonary valve stenosis (PS), and combined defects of atrial and ventricular septum or TOF are not uncommon [8].

Heart failure (HF) secondary to CHD is a clinical syndrome, which can also be confused with non-cardiac causes. The aetiology of HF differs greatly between children and adults. Congestive HF in children with CHD is usually not caused by pump dysfunction, but is characterized by hemodynamic disturbances due to left-to-right shunts, pulmonary over-circulation, and volume overload causing pulmonary oedema, manifesting as increased respiratory effort and tachypnoea, recurrent pneumonia, poor feeding or even generalized oedema or hydrops fetalis [9].

CHD can present as HF in the first week of life in duct dependent lesions. Few other CHDs may present around 6-8 weeks of life or around two years of life when the pulmonary pressures further decreases. Others may remain asymptomatic until adulthood. [5]

Medical management of CHD varies depending on the type of lesion. Prostaglandin infusion is used in a duct dependent lesion. Propranolol reduce risk of cyanotic spells in TOF patients and varies types of anti-failures can be used for CHD patients with HF. [5]

Diuretics, calcium channel blockers, beta blockers and less commonly the digitalis are used worldwide and in Fiji. Diuretics are the first line of therapy for diastolic dysfunction. Diuretics reduce pulmonary congestion and relieve symptoms such as orthopnoea, cough and dyspnoea. Injudicious or excessive use will reduce preload and result in diminished cardiac output [8, 9].

Corrective cardiac surgery in early infancy seems to be the most effective therapy to reverse neurohormonal activation in patients with CHD. However, infants with complex cardiac anomalies frequently may not undergo a complete repair in infancy or may need palliative surgery like pulmonary artery banding or aortic-pulmonary shunts which may have a negative impact on their prognosis. [9] Rapid advances have taken place in the diagnosis and treatment of CHD. There are diagnostic tools available, upon trained hands accurate diagnosis can be made even before birth. With current treatment modalities, more than 75% of infants born with critical CHD can survive beyond the first year of live [1].

In developed countries like United States of America, (USA) yearly 32,000 infants are born with CHD; however mortality from heart defects has declined, although CHD remains a major cause of death in infancy and childhood. Age at death is increasing, suggesting that more affected persons are living into adolescence and adulthood [10, 11].

Spectrum of CHD is not race related. A study in Korea showed a similar spectrum of CHD to USA in that population. Despite CHDs like PDA being higher in preterm babies, the incidence of CHD is higher in full term neonates as interestingly documented by the Korean study [8]. The most common symptom that would lead to an echo study was auscultation of a murmur [7].

A retrospective descriptive study in Kanpaur, India, by Koopar and Gupta identified a very high prevalence of CHD, of 26/1000 patient [13]. The reason for this high incidence compared to international rates and previous studies is that this study looked at CHD in 0-15 year's age group and included mild, moderate and severe forms of CHD. Other studies looked at 0-5 years or 5-15 years groups and did not include mild CHD. Similarly this study will include all cases up to 15 years of age. However, it will exclude the mild CHDs (PDAs, VSDs, and ASDs) which spontaneously close by 2 months of age; the age at which a repeat echo would be done for the neonates screened at birth for various reasons and a mild CHD is found, at CWMH.

There is no published data on incidence, prevalence, spectrum or outcome of CHD in the Pacific in the last 15 years. Unpublished data by Mataika R, 2012. [12], a study on the burden of CHD at CWMH, estimated the incidence of CHD in CWMH at 4.9/1000 live births.

All the Pacific island nations rely on overseas cardiothoracic surgeons or FIFO cardiac teams to do consultation and surgery for the children with CHD. Papua New Guinea with a population of more than a million and about 100 children needing surgery each year have taken steps into training cardiac surgeons. Under the leadership of Open Heart International, the local team had performed their first cardiac surgery with the visiting team present in the operating room in 2016 [14].

Unfortunately, the privileges of early diagnosis and early management available to children in developed countries are currently inaccessible to the majority of children in developing countries like Fiji, afflicted with CHD, leading to high morbidity and mortality.

METHODOLOGY

This is a descriptive, retrospective, single-centred study at CWMH. Names of children with CHD were collected from various hospital registries. Folders were retrieved, and if cases fulfilled the inclusion criteria and not the exclusion criteria, they were coded with a unique identification number. Using the unique de-identified numbers (UDN), details from the folders were entered into an excel spreadsheet. Excel data analysis and chi square tests was used to analyze the data.

Inclusion criteria:

- All children <15 years old with newly diagnosed CHD confirmed by echo between January 1st 2012 to the December 31st 2016.
- All children <15 years old who died and CHD was confirmed by post mortem (PM) between 1st January 2012 to 31st of December 2016.

Exclusion criteria:

- No documentation of echo or PM findings noted in the folder.
- Those with spontaneous closure of VSD/ASD or PDA confirmed by a repeat echo at 2 months of age.
- Children referred from other divisional hospitals or Pacific islands.

Process for Data Collection

A search for CHD cases was performed using the Patient Information System (PATIS), ward admission registries, echo registry, FIFO team reports and consultant clinic cardiac patients' records.

There was 89% folder retrieval rate. The Principle investigator (PI) looked through each of the folders and included or excluded each case as appropriate. Each case in the sample group was then assigned a unique unidentified number.

Process for Data Management and Analysis

The coded data collection form (DCF) was used to record information from the folders. De-identified data from the DCFs were entered into an excel spread sheet. Only the PI had access to list of the patients' names corresponding to UDN.

Ethical Considerations

Confidentiality was maintained throughout as discussed above. This study was approved by Fiji National University College Health Research Ethics Committee (CHREC) and Fiji National Health Research and Ethics Committee (FNHREC) of MOH, Fiji.

Pretest

The DCF was pretested with 10 folders of CHD cases and adjustments were made accordingly prior to data collection.

RESULTS

In this five-year study, 348 cases were identified, 311 folders were retrieved, giving this study a folder retrieval rate of 89%. Ninety-nine cases were excluded as they did not fulfil the inclusion criteria or they fulfilled the exclusion criteria. [Figure 1]

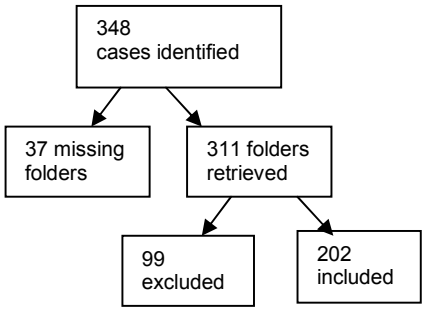

Figure 1: Total cases identified and excluded

The incidence of CHD in CE division in this study is 0.6% (5.8/1000) live births. If the missing folders cases were added to the sample size, the incidence of CHD would have increased to 0.7% (7/1000).

There was no difference in the incidence of CHD between males (51%) and females (49%). However the incidence of CHD in the FoI was double that of the i-Taukei population. (OR of 2 and $p < 0.0001$) The most common acyanotic CHD is VSD and the most common type of cyanotic CHD is TOF. [Figure 2]

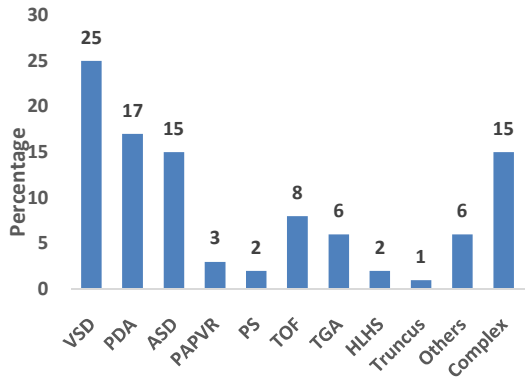

Figure 2 : Types of Congenital Heart Disease by proportion

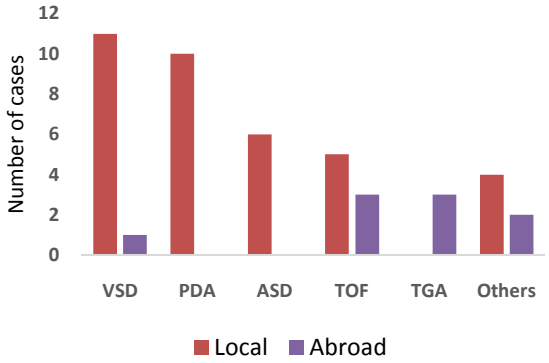

Figure 3: Congenital Heart Diseases cases that underwent surgery

The most common indication for echo referral was a cardiac murmur, followed by cyanosis and tachypnoea. Syndromic neonates also contributed a significant number of cases referred for echo, as part of the syndromic neonates' workup. The majority of cases (82%) were diagnosed before 1 month of age.

There was no statistically significant association in the outcome (death or alive) comparing the age at diagnosis for the critical CHD cases. (Comparing diagnosis <72 hours & >72 hours); P value of 1.

The three most common causes of morbidity (admissions to CWMH) were pneumonia, congestive cardiac failure and failure to thrive. Other reasons were infective endocarditis (IE), cyanotic spells and others.

Forty-six cases (23%) had surgery within the study period. All cases selected for surgery by the three FIFO cardiac teams within the period of study required only one-stage corrective cardiac surgery. Nine cases were considered too high risk and referred for surgery abroad. [Figure 3]

One case out of the 46 cases needed a redo surgery and there was one death. Hence the survival rate for the surgeries done locally was 98% and survival rate for cases done abroad was 100%.

Seventy-six (39%) children in the cohort died. Ninety-five percent of them died before their first birthday. The majority (65%) of these cases were acyanotic CHD cases, and 20% of them had PDA with complications.

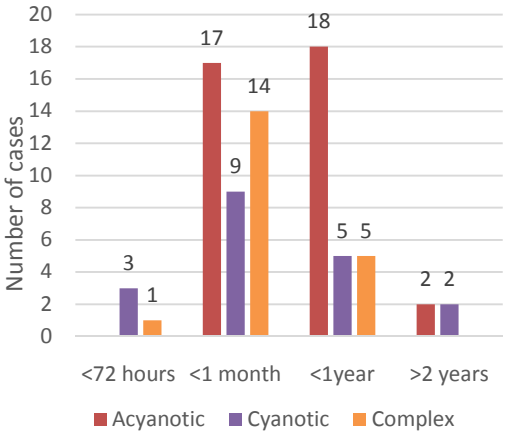

Figure 4: Age at death by Type of Congenital Heart Disease

DISCUSSION

The incidence of CHD in CE division in this study is 0.6% (5.8 per 1,000) live births. This is similar to Mataika's study in 2012 with an incidence of 0.5% (4.9/1000) live births [12]. If the missing folders cases were included, the incidence would have increased to 0.7%. This number would have further increased if positive post-mortems for CHD cases presented as Dead on Arrival (DOA) to other medical facilities within the CE division were included in the sample but this data was not available. It should also be noted that no routine antenatal ultra sound scan screening for CHD was made antenatally, nor was post natal screening with pulse oximetry for critical CHD performed. Cases were referred for echo from within CWMH or the periphery (public health) if clinically indicated. Some CHD cases lesions with subtle findings may not be detected such as ASDs which could account for this lower incidence rate compared to the worldwide incidence of 1% (10/1000) live births [2,5,6,7,8].

There was no difference in the incidence between genders. This is a similar finding to the Korean study [8]. Interestingly, when looking at raw numbers, the i-Taukei population constitute 68% of the cases with CHD compared to the FoI (34%). However, i-Taukei women delivered 78% of the birth cohort, compared to FoI women (17%). Hence the incidence of CHD in the FoI population is twice the incidence in i-Taukei children. (OR of 2, $p< 0.0001$) This could be attributed to the health seeking behaviour of the FoI parents compared to the i-Taukei parents. A similar study in Central Australia comparing the incidence in the Aboriginal and non-Aboriginal population did not find any difference in the incidence between the 2 groups [3].

VSD was the most common acyanotic CHD, followed by ASD and PDA. TOF was the most common cyanotic CHD, followed by Transposition of the Arteries. This is similar findings to the Australian and Indian study [3, 11]. In the Korean study [8], the most common lesion was ASD, and in Mataika's unpublished study [12], TGA was the most common cyanotic CHD. Complex CHD made up 15% of the cases. Complex CHD are cases that can have a mixture of acyanotic and cyanotic CHD and do not fit into any particular type of CHD. These cases were only offered palliative care in Fiji, which does not include palliative surgery.

The 46 cases that had surgery; single repairs were done locally for simple defects such as VSD, ASD and PDA, PS. All three TGA cases (25% of all TGA cases) that had surgery were performed abroad. Seventy-five percent of the TGA cases died, due to late diagnosis, unavailability of prostaglandin or funding for urgent transfer for corrective surgery abroad. Of the 8 TOF cases that had repair ocally or abroad, all had a single-stage repair. Children requiring multiple-stage operations were not operated for a range of reasons, mainly due to lack of funding for multiple surgeries for the TOFs with critical or Severe PS, the need for pacing and management of complications arising from staged operations.

Unlike Mataika's study [12] where the majority of cases had surgery abroad, and most of the mortality was among children with cyanotic CHD, in this study, only 9 (~20%) of the cases had surgery abroad. This is mainly due to the ceasing of major sponsor of funding by Vodafone ATH Foundation (England) 2007-2011.

Twenty-six percent of the deaths were attributable to complex CHD. Palliative surgery cannot be supported locally and the MOH does not fund palliative surgery abroad. However, the majority of deaths were acyanotic CHD (~ 50%) and 65% of these were VSDs, ASDs and PDAs cases who required anti-failure medications. This reflects mortality due to the absence of corrective surgery locally.

The outcome of critical CHD cases, excluding the complex CHD cases (since no palliative surgery was offered) is statistically not significant if diagnosed early (<72hours) or late (>72 hours) This is expected, as unlike other countries where early detection would mean early corrective surgeries, in Fiji, the process of transferring children abroad has

many challenges. These include unavailability of prostaglandins, tele-medicine challenges (image transfer) for confirmation/acceptance, and most importantly the lack of funding or insurance cover.

Despite there being three FIFO cardiac teams visiting Fiji annually, there is still at least a six months gap between visits. During that period, CHD cases diagnosed and requiring urgent surgery were managed medically, until a cardiac team visits. However many of these cases do not survive as they need corrective surgery.

The majority of children who had surgery both locally and abroad are well and are expected to have a normal life expectancy for Fiji. Within 6 months post-surgery, one case required a redo surgery and one child died. That brought the survival rate of local surgeries to 98%, compared to 100% for the surgeries done abroad in New Zealand and India.

LIMITATIONS OF THE STUDY

There was inaccurate recording of the echo studies in the echocardiography registry book. Many critically-ill children had portable echocardiography studies that were not recorded in the register. Other cases of CHD may have been missed because the folders were not correctly or fully classified.

Data was collected from the folders or admissions and hence only admitted cases that presented to CWMH were included in this study. Cases that presented to other medical facilities within the Central/Eastern division and had CHD confirmed by PM were not included in this study.

There are about 80 to 100 cases of intrauterine deaths yearly at CWMH. Post-mortems are not performed routinely in Fiji to determine the cause of death and from published literature this would add 8 to 10 CHD each year.

CONCLUSION

CHD continues to cause high morbidity and contributes significantly to the neonatal and infant mortality rate in Fiji.

While there is a need to continue to improve early detection of CHD to elucidate the burden of disease. Fiji needs to strengthen its management/intervention strategies, especially the unmet need for corrective surgeries, and in the interim, securing funding for urgent referrals, and making longer term plans for the establishment of a Paediatric cardiac-surgical unit in the medium term (five to ten years).

Fiji has been dependent on FIFO teams for more than 20 years and needs to invest in the human resource and infrastructure needed to provide cardiac surgical services. Once Fiji has the infrastructure, resources and local trained human resources, improved methods of early detection like antenatal foetal anomaly scanning and screening for critical CHD postnatally with pulse oximetry, Fiji can then truly address CHD to reduce morbidity and mortality.

RECOMMENDATIONS

The recommendations made from this study are:

1. Establish a national standard for the diagnostic requirements for congenital heart disease including personnel, criteria and equipment.

2. Establish a national database for CHD. This can be piloted at CWMH as there is constant/regular echo clinic done at CWMH.

3. The unmet need for curative corrective surgery for CHD needs to be addressed acutely by strengthening the private-public partnerships through NGO's like Children's Heart Foundation in securing funds for cases that need urgent cardiac referrals abroad.

4. Consider a long term plan, to establish Paediatric Cardiac surgical unit in the next 5 to 10 years. A

local team to be trained in echocardiography, Paediatric cardiology, Paediatric cardiac surgeons and other cadres needed for such a highly specialized unit.

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