East African Medical Journal Vol. 84 No. 7 July 2007
MANAGEMENT PATHWAY FOR CONGENITAL HEART DISEASE AT KENYATTA NATIONAL HOSPITAL, NAIROBI
M.N. Awori, MBChB, MMed (Surg), Tutorial Fellow, S.W.O. Ogendo, MBChB, MMed (Surg), Associate Professor, S.W. Gitome, 5th-year Medical Student, S.K. Ong'uti, 5th-year Medical Student and N.G. Obonyo, 3rd-year Medical Student, Department of Surgery, College of Health Sciences, University of Nairobi, P.O. Box 2483-00202, Nairobi, Kenya

Request for reprints to: Dr. M.N. Awori, Department of Surgery, College of Health Sciences, University of Nairobi, P.O. Box 2483-00202, Nairobi, Kenya

MANAGEMENT PATHWAY FOR CONGENITAL HEART DISEASE AT KENYATTA NATIONAL HOSPITAL, NAIROBI

M.N. AWORI, S.W.O. OGENDO, S.W. GITOME, S.K. ONG'UTI and N.G. OBONYO

ABSTRACT

Background: **Congenital heart disease (CHD) is a significant cause of death amongst infants. The timing of treatment in relation to the natural history of the disease correlates with the treatment outcome.**

Objectives: **To determine the age at first suspicion of CHD, the age at confirmation of the diagnosis of CHD and the percentage follow-up at the first post diagnosis out patient clinic and to determine the influence of patient's sex, parental income and parental education have on the MP.**

Design: **A five year retrospective study.**

Setting: **Kenyatta National Hospital between January 1st 2000 and December 31st 2004.**

Subjects: **Two hundred and fourteen patients were studied.**

Results: **The overall mean age at referral to a paediatric cardiologist was 16.9 ± 24.4 months [n = 102]. The mean age at which CHD was confirmed by echocardiography was 18.6 ± 25.6 months [n = 202]. The mean age at which CHD was first suspected in patients from the province with the highest parental income was 9.5±5.1 months [n = 6]. The mean age at which CHD was first suspected in patients from a province with a significantly lower parent income was 19.1 ± 23.2 months [n = 22], (p = 0. 046). The mean age at which CHD was confirmed in referred male patients was 16.0 ± 17.6 months [n=48] and the mean age at which CHD was confirmed in referred female patients was 18.8 ± 21.7 months [n = 52] (p = 0.25).**

Conclusion: **The mean age at referral to a paediatric cardiologist was 16.9 months. This suggests that a significant number of patients may miss the opportunity to have optimal surgical intervention. Parental income appears to influence the MP, however, the level of parental education and patient sex did not.**

INTRODUCTION

Congenital heart disease (CHD) comprises a group of disorders of the heart and great vessels that results from aberrations of normal organogenesis. CHD is a significant cause of death amongst infants (1). In the developing world, the majority of children born with CHD do not receive medical attention (2). The timing of treatment in relation to the natural history of the disease is of utmost importance as it often correlates with the outcome of treatment.

The incidence of CHD has been shown to be constant worldwide (3). British literature estimates the incidence of CHD to be approximately 8:1000 live births (4). The incidence of CHD in Tanzania was found to be 7:1000 live births (5).

The current population in Kenya is 32.2 million, the crude birth rate is 37.5 : 1000 and the infant

mortality rate is 77: 1000 (6). Computations based on these data and on knowledge of the natural history of CHD suggest that approximately 6000 children are born in Kenya every year who require surgery involving cardiopulmonary bypass.

Some newborns have obvious symptoms and signs of heart disease; others may have complex lesions that are not detected for months. Routine neonatal examination fails to detect more than half of the neonates with heart disease (7). The specificity of the symptoms and signs usually is not adequate for planning definitive therapy. Congenital heart diseased may be suspected if a patient has a history of feeding difficulty, cyanosis, or frequent chest infections. The presence of murmurs, signs of cardiac failure, cyanosis or low anthropometric indices may also be useful clues to the presence of CHD (8). Once a clinical abnormality suggestive of CHD is recognised, an investigation must be carried out until a definitive diagnosis is established. Two-dimensional colour flow doppler echocardiography (ECHO) has become the preferred initial assessment once CHD is suspected on clinical grounds and has in many cases eliminated the need to perform cardiac catheterisation. Generally speaking, ECHO has a sensitivity of about 90% and a specificity of up to 100% (9). In most cases, the decision to operate can be based on ECHO findings alone. Cardiac catheterisation is still indicated for certain cases where it is important to accurately measure intracardiac chamber pressures prior to surgery.

Most diagnosis of CHD are made after birth, however, foetal echocardiography may detect the presence of an anomaly and in many cases may establish a specific diagnosis. Prenatal diagnosis of CHD is important for proper perinatal management. Antenatal diagnosis is crucial as special obstetric and neonatal management may be required in which case delivery at a tertiary care centre will be advantageous (10).

The worldwide data base on outcomes of interventions for CHD is still developing, in view of this, long term follow-up in the out-patient clinic is desirable as this will aid in the development of the outcome data base. This follow-up will have two main effects. Firstly, it will enable healthcare providers to deliver timely treatment for complications. Secondly it will enable healthcare providers to assess the quality of their care and thereby provide avenues for health care improvement.

The management of CHD consists of three phases: diagnosis, treatment and follow-up. For simplicity these three phases will be referred to as the MP. An assessment of the efficacy and efficiency of each phase of the MP should provide a means of determining the overall performance of the MP.

The typical MP usually involves the following steps:

(i) Clinical suspicion (by parents or health care workers) of CHD
(ii) Confirmation of CHD by ECHO (possibly cardiac catheterisation)
(iii) Surgical intervention
(iv) Follow-up

The ideal MP exists when the four steps outlined above are carried out in timely fashion; the time frame varies according to the nature of the lesion. The aim of surgical intervention is to correct or palliate the lesion before the natural progression of the disease renders intervention impossible or less effective. Delays along the MP translate into an increased morbidity and mortality.

By assessing the current status of the management of CHD at a leading national health institution, proponents of optimal care for patients with CHD will have a framework upon which to develop protocols for improvement in care. No study has been done to objectively ascertain the standard of care given to patients with CHD locally.

The objective of the current study was:

(a) to determine the ages at first suspicion of CHD, to determine the ages at confirmation of the diagnosis of CHD and to determine the percentage follow-up at the first post-diagnosis out patient clinic.
(b) To determine the influence patient sex, parental income and parental education have on the status of the MP for CHD.

MATERIALS AND METHODS

This was a five-year retrospective descriptive study conducted between January 1st 2000 and December 31st 2004. The study took place at KNH (Nairobi). All patients younger than 12 years of age seen at KNH and who subsequently had a diagnosis of CHD confirmed by echocardiography were included in the study.

All patients seen at KNH with a diagnosis other than CHD and all patients seen at KNH with an unconfirmed diagnosis of CHD were excluded from the study.

A sample was taken of all patients younger than 12 years of age seen at KNH, and who subsequently had a diagnosis of CHD confirmed by ECHO. The investigators examined patient case notes retrieved by the records clerk and relevant data found therein was entered into an individualised data sheet. Surrogate data on parental income and parental education (according to province) was obtained from a contemporaneous demographic and health survey (6). The surrogate for the level of parental income was considered to be the percentage of adult males in the occupation with the highest wealth quintile. The surrogate for the level of parental education was considered to be the percentage of adult females with no access to any level of formal education and the percentage of adult females with no access to mass media. Data were summarised using graphs and charts. Ratios (percentages) were used to summarise discrete statistics, means (central tendency) and standard deviation (spread) were used for continuous variables. Microsoft Excel was used to calculate means and standard deviation. The student t-test for differences in two sample population means was used to look for significant differences in mean ages, the Chi-square test was used to assess differences in population characteristics reported in the demographic and health survey (6) ($p<0.05$ was considered to be significant).

The study was embarked upon following approval by the Kenyatta National Hospital Ethics and Research Committee.

RESULTS

A total of 214 patients were included in the study. The overall mean age at which CHD was first suspected was 16.9 ± 24.4 months (n = 190), compared to 15.8 ± 21.2 months (n = 90) and 17.8 ± 27 months (n = 100) for patients referred to KNH and those presenting directly to KNH respectively (p = 0.138).

The overall mean age at which CHD was confirmed by ECHO was 18.6 ± 25.6 months (n = 202). The mean age at which CHD was confirmed in patients referred to KNH and patients presenting directly to KNH was 17.9 ± 22.2 months (n= 100) and 18.6 ± 27.7 months (n = 102) respectively (p = 0. 16).

The mean duration from echocardiogram request to echocardiogram performance (ECHO interval) was 26.3 days (SD 55.6 days; 192 had adequate data and were included in this calculation). Of the 12 patients who required a Modified Blalock-Taussig shunt (MBTS) following ECHO findings, six eventually received it. The mean duration from the performance the echocardiogram confirming the need for a modified BT shunt to the surgical creation of the MBTS was 8.0 ± 11.8 months. Of the 164 patients who had data regarding follow-up, 88 (53.7%) were completely lost to follow-up after discharge from their last admission to KNH despite being scheduled for a paediatric outpatient clinic (POPC) visit.

The surrogate value for parental income in Coast province is significantly higher than in Eastern province (Chi-square test applied to demographic survey data) (6). The surrogate value for the level of parental education is lower in Coast province than in Eastern Province. Eastern and Central provinces have similar levels of surrogate parental income but significantly different levels of surrogate parental education (Tables 1 to 3).

The place where the diagnosis of CHD was first suspected is shown in Figure 1. The place where the diagnosis of CHD was confirmed is shown in Figure 2.

Table 1

Age CHD suspected: Coast compared to Eastern Province

Province	Age CHD suspected	P-value
Coast	9.5 ± 5.1 months (n = 6)	0.046
Eastern	19.1 ± 23.1 months (n = 22)	

Table 2

Age CHD confirmed: Coast compared to Eastern Province

Province	Age CHD confirmed	P-valve
Coast	18.0 ± 13.9 months (n = 6)	0.4
Eastern	19.7 ± 23.1 months (n = 22)	

Table 3

Age CHD suspected: Central compared to Eastern Province

Province	Age CHD suspected	P-value
Central	17.5 ± 23.5 months (n = 25)	0.309
Eastern	19.1 ± 23.1 months (n = 22)	

Table 4

Age at confirmed diagnosis: comparison between sex in referred cases

Sex	Age CHD confirmed	P-value
Male	16.0 ± 17.6 months (n = 48)	0.25
Female	18.0 ± 21.7 months (n = 52)	

Figure 1

Place diagnosis of CHD suspected

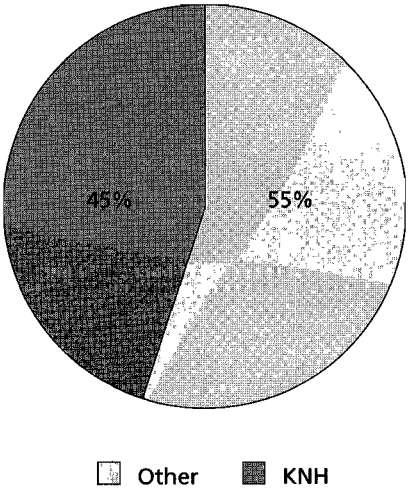
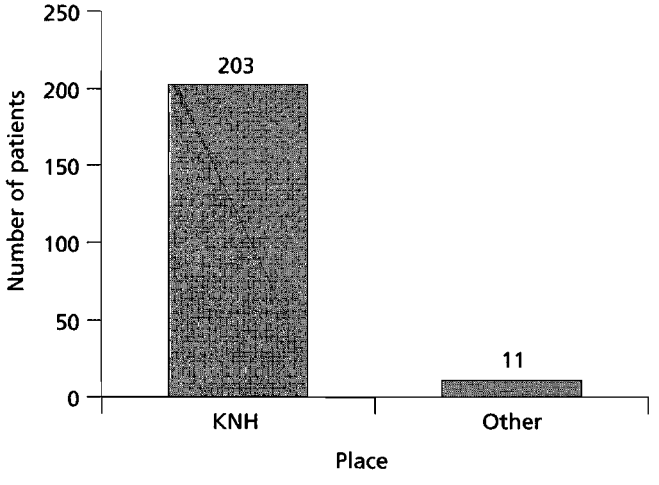


Figure 2

Place diagnosis of CHD confirmed



DISCUSSION

Irreversible changes to the pulmonary vasculature that preclude corrective surgery in children with CHD may occur by six months of age. The confirmation of CHD by ECHO is usually carried out by a paediatric cardiologist. The timing of referral for paediatric cardiac care is a crucial step in the MP. The mean age at referral to a paediatric cardiologist in one developed country was 1.9 months (11). In some regions each successive step in the MP occurs in different geographical locations separated by considerable distances (12). This makes it more challenging (from a temporal perspective) to achieve ideal MP's in these regions.

Kumar suggests that the two biggest contributors to deficiencies in MP's in developing nations are poverty and ignorance (2). He states that paediatric cardiac care is too expensive for the average Indian family. Kumar argues further that ignorance manifests itself as a lack of awareness of the need (on the part of healthcare providers) for urgent referral of patients with CHD to a paediatric cardiac care programme.

Over 60% of children in this study locality present with lesions that cause irreversible pulmonary vascular changes. The mean age at referral in our study was 16.9 months. This figure suggests that a significant number of patients may be missing the window of opportunity for surgical intervention.

The overall mean age at which CHD is first suspected gives a national perspective with respect to the management of CHD. By separating these patients into two groups: (a) The mean age at which CHD is suspected for the first time in a health care facility other than KNH (referred patients) and (b) The mean age at which CHD was suspected for the first time at KNH (KNH patients), we create an opportunity to examine the efficiency of the referral mechanism with respect to CHD.

The mean age at which CHD was first suspected in referred patients and the mean age at which CHD was first suspected in KNH patients is not significantly different. These findings suggest that there is no significant delay in referring patients with suspected CHD to KNH. This is a pleasant surprise as the anecdotal evidence is replete that

the local referral system has collapsed. However, CHD is first suspected and subsequently confirmed much later in our locality compared to some developed nations. Kumar' s theory that medical care provider ignorance contributed to delays in referral and subsequent treatment is mitigated by the fact the there is no significant difference in the age CHD is confirmed in patients referred to KNH and in patients first seen at KNH. It appears more likely that a lack of adequate access to health care providers contributed to the late diagnosis of CHD locally. The reasons for this poor access to health care providers may be financial (patients parents can not afford to see the doctor), geographical (it is difficult to physically travel to the doctor) or even related to human resource issues (to few doctors).

Patients from Coast Province were suspected of having CHD at a significantly earlier age than patients from Eastern Province. It would appear that the level of parental income influences the MP at the "suspicion of CHD" level and that the level of parental education may not influence the MP at all. This argument is supported by the finding that the age at which CHD was first suspected in children from Eastern province and Central province was not significantly different.

It is interesting to note that the age at which CHD was confirmed in Coast and Eastern provinces was not significantly different. This finding suggests that the level of parental income has no influence on the MP at the "confirmation of CHD" level. This apparent lack of influence might be explained by a shortage in ECHO services in the country. In this situation a lack of parental finances would cease to be a bottle-neck in the MP, instead, a lack of ECHO services would be the bottle-neck. If this is indeed the case, this finding could be an indicator that the technical arm of our local health care service may require upgrading.

A study carried out on an Indian community found that there was a gender bias favouring the treatment of male children (13). Patient's sex does not appear to influence the MP locally as there is no significant difference in the age at which CHD was confirmed in referred male and female patients (Table 4).

The mean age at which CHD was confirmed was 18.6 months and the ECHO interval was 26.3 days. These figures suggest that there may be a significant delay (1.9 months) from the time CHD is first suspected to the time confirmatory ECHO is requested. This delay could increase local mortality rates. The 26.3 days ECHO interval would certainly prove fatal for certain types of Transposition of Great Arteries and would render optimal corrective surgery impossible in others. This prolonged ECHO interval could have economic implications. It is not unusual to come across mothers and sick infants lying in a ward bed simply waiting for an ECHO to be done. This is an unnecessary lodging cost that stretches the already meager resources.

Almost half of all cases of CHD seen were first suspected of having CHD in KNH. Considering that KNH serves Nairobi residents in a non-referral capacity, and that the population of Nairobi is estimated at three million, it is surprising to note that the balance of the population of Kenya (approximately 32.2 million) appears to have yielded a similar number of CHD patients (via nation wide referrals) as Nairobi. This finding could suggest two things. Firstly, there may be a problem with the referral chain. Our finding that there was no significant difference in the age at first CHD suspicion and the age at CHD confirmation between KNH patients and referred patients argues against this alternative. The second suggestion is that there could be a significant number of patients out side the Nairobi area that never come into contact with medical personnel and so never enter the referral chain. This seems to be the more likely scenario and if this indeed is the case, it is a grave problem as it implies that a large number of children with CHD do not receive medical attention.

It has sometimes been said that KNH is a dumping ground for patients once management can no longer proceed in the private sector on financial grounds. The results of this study suggest that KNH provides a very real diagnostic role as most cases of CHD were confirmed in KNH. This strengthens the case for allocating resources to improve the service provided by KNH.

The small number of patients actually receiving the required surgery precluded any meaningful analysis of the effect of parental income; parental education and patient's sex might have on this stage of the MP. However, the small percentage of patients actually receiving the required surgery and the protracted surgical interval (as seen in patients requiring MBTS) seem to suggest that the surgical phase of the local CHD-MP needs to be developed.

There also seems to be a problem with the outpatient follow up as more than 50% of patients are completely lost to follow-up after their last discharge from KNH. It was not possible to determine the percentage attendance of the first post diagnosis out patient clinic. However, the percentage attendance of scheduled outpatient clinics after the patients last recorded discharge from KNH gives some idea of the level of patient follow-up. This high out patient clinic default rate may be causing another grave problem as a significant number of patients are discharged from KNH with the instructions that the diagnostic ECHO be done as an out-patient. This means that we cannot determine the fate of a significant number of children who were presented to our health care system by parents seeking our help.

In conclusion, the results of this study suggest that a significant number of neonates and infants with CHD miss the window of opportunity to have optimal surgical intervention. The level of parental income appears to have an influence on the MP of CHD locally. Patient's sex and the level of parental education appear to have no influence on the MP. It is evident that there are problems at every level of the MP of CHD locally. Further investigation is required to determine how best the CHD-MP in this region may be optimised.

REFERENCES

1. Schwartz S.I. 1998. Principles of surgery. 7th international edition. McGraw Hill: 792.
2. Kumar R. Congenital heart disease management in developing countries. *Pediatr. Cardiol.* 2003; **24:** 311.
3. Abdulla R. What is the prevalence of congenital heart diseases? *Pediatr. Cardiol.* 1997; **18:** 268.
4. Anderson J.R. and Hunt I. Bailey and Love's short practice of surgery, 24th international students' edition. *Arnold.* 2004; 905.
5. Padmini R.A. and D'Souza N.Y. The incidence and pattern of congenital heart defects in Tanzanian children. *East Afr. Med. J.* 1976; **53:** 36.
6. Central Bureau of Statistics (CBS), Kenya, Ministry of Health (MOH), Kenya and ORC Macro. 2004. Kenya Demographic and Health survey 2003. Calverton, Maryland: CBS, MOH, and ORC Macro.
7. Wren C., Richmond S. and Donaldson, L. Presentation of congenital heart disease in infancy: Implications for routine examination. *Arch. Dis. Child Fetal Neonatal Ed.* 1999; **80:** 49-53.
8. Dimiti A.I. and Anabwani G.M. Anthropometric measurements in children with congenital heart disease at Kenyatta National Hospital (1985-1986). *East Afr. Med. J.* 1991; **68:** 757-764.
9. Gutgesell H.P., Huhta J.C., Latson L.A., *et al.* Accuracy of two-dimensional echocardiography in the diagnosis of congenital heart disease. *Amer. J. Cardiol.* 1985; **55:** 514-518.
10. Strauss A., Toth B., Schwab B., *et al.* Prenatal diagnosis of congenital heart disease and neonatal outcome-a six year experience. *Eur. J. Med. Res.* 2001; **6:** 66-70.
11. Fixler D.E., Pastor P., Sigman E., *et al.* Ethnicity and socioeconomic status: Impact on diagnosis of congenital heart disease. *J. Amer. Coll. Cardiol.* 1993; **21:** 1722-1726.
12. Tefuarani N., Hawker R., Vince J., *et al.* Congenital heart disease in Papua New Guinean children. *Ann. Trop. Paediatr.* 2001; **21:** 285-292.
13. Amin M. and Khondoker F. A contingent valuation study to estimate the parental willingness-to-pay for childhood dairrhoea and gender bias among rural households in India. *Health Research Policy and Systems.* 2004; **2:** 3.