Vol. 16(3), pp. 22-26, May 2024
DOI: 10.5897/AATCVS2024.0066
Article Number: 9F07C5C72112
ISSN 1994-7461

http://www.academicjournals.org/AATCVS

ACADEMIC JOURNALS
expand your knowledge

African Annals of Thoracic and Cardiovascular Surgery

Case Report

Current status of the paediatric congenital heart disease management pathway in Kenya

Mark Nelson Awori[1]*, Daniel Ojuka[1], Diana Marangu[2] and Paul Bannon[3]

[1]Department of Surgery, School of Medicine, University of Nairobi, P. O. Box 19676-00202, Nairobi, Kenya.
[2]Department of Paediatrics and Child health, School of Medicine, University of Nairobi, P. O. Box 19676-00202, Nairobi, Kenya.
[3]Department of Cardiothoracic Surgery, University of Sydney, NSW 2006, Australia.

Received 11 March, 2024; Accepted 3 April, 2024

The objective of the current study was to assess the current status of the congenital heart disease (CHD) Management Pathway (MP) in the same public hospital in Kenya, following an assessment conducted sixteen years ago that found all phases of the MP to be inadequate. Over the course of one year, patients under 12 years of age with congenital heart disease (CHD) were consecutively recruited from the general pediatric wards at Kenyatta National Hospital (KNH) in Nairobi. All recruited patients required intervention, either through surgery or cardiac catheterization, and were subsequently followed up for one-year post-recruitment. Data collected encompassed the age at which CHD was first suspected, age at diagnosis, age at intervention, and the outcome at the one-year follow-up. Out of the initial 49 patients recruited, 47 were successfully contacted via mobile phone at the one-year mark post-recruitment. The mean ages at which congenital heart disease (CHD) was first suspected and diagnosed were 35.3 weeks (SD +/- 91.1) and 43.4 weeks (SD +/- 97) respectively, with the mean age at intervention being 47.5 weeks (SD +/- 51). Thirty-five percent of all patients underwent surgery, and 96% were successfully followed up. It is possible that over the last 16 years, there has been an improvement in at least two phases of the Management Pathway (MP). However, the proportion of patients receiving surgery has remained unchanged, suggesting a need for future resource allocation to this phase.

Key words: Congenita,l cardiac, pathway, diagnosis, treatment, follow-up.

INTRODUCTION

This study examined the current status of the Management Pathway (MP) at the same public hospital where it was previously assessed 16 years ago, with regards to the management of congenital heart disease (CHD). Congenital heart disease (CHD) is characterized by congenital malformations of the heart and/or great vessels, with a global prevalence of 10 per 1000 live births (Liu et al., 2019). Twenty-five percent of patients are diagnosed with 'critical' congenital heart disease (CCHD), necessitating intervention (surgery/cardiac catheterization) within the first year of life for survival (Glidewell et al., 2015; Samanek et al., 1992). Patients with haemodynamically significant non-critical CHD who do not receive timely intervention often progress to 'inoperability' (Gan et al., 2014). The age at diagnosis is a critical factor in the MP of CHD, which consists of three

*Corresponding author. E-mail: mnawori@yahoo.com. Tel: +254707366336

Table 1. Evidence based standards of care (Park, 2014).

Lesion	Treatment	Age by which treatment should be delivered
Simple transposition of great arteries (TGA)	Arterial switch operation (ASO)	1-3 weeks of age
Obstructed Total anomalous pulmonary venous drainage (TAPVD)	Repair	1-4 weeks of age
Hypoplastic left heart syndrome	Norwood procedure	1-4 weeks of age
Correctable shunt lesions with pulmonary over circulation and pulmonary hypertension	Repair	By 3 months of age
Single ventricle lesions with pulmonary over-circulation and pulmonary hypertension	Pulmonary artery banding	By 3 months of age
Cyanotic neonates/infants (arterial oxygen saturation <75%)	Central shunt (CS) or modified Blalock-Taussig shunt (MBTS)	1week to 5 months of age
TGA with Ventricular septal defect and pulmonary valve stenosis	ASO OR REV procedure	By 6 months of age
Cyanosed neonates/infants correctable lesions with; arterial oxygen saturation >75%; no or mild pulmonary hypertension	Repair	By 6 months of age
Single ventricle with normal pulmonary tree	Bi-directional Glenn shunt	By 4-8 months of age
Single ventricle with normal pulmonary tree	Fontan connection	At 18 months to 4 years of age
Simple coarctation of the aorta	Repair	At 2 to 4 years of age
Aortic valve stenosis	Repair	At 10 years of age
Small Shunt lesion with normal sized heart and normal pulmonary artery pressure	Surgery not indicated	Not applicable

REV=Reparation a l'Etage Ventriculaire.

phases: diagnosis, treatment, and follow-up, serving as a pragmatic approach to healthcare delivery for CHD patients (Awori et al., 2007).

Since the inception of cardiac surgery, ongoing research has provided evidence-based standards for optimal care (OC) of CHD patients, as summarized by Park (2014). OC entails delivering all three phases of the MP in accordance with these evidence-based standards. Sixteen years ago, Awori et al. (2007) identified significant deficiencies in all phases of the MP at a public hospital in a low-middle-income country (LMIC). Considering changes in Kenya's economic status (Fleming, 2022; World Bank, 2023) and government structure (Kimenyi, 2013) over the past 13 years, these alterations may have impacted the MP, prompting the need for reassessment 16 years ago. Table 1 shows the evidence-based standards of care (Park, 2014).

MATERIALS AND METHODS

A prospective cohort study conducted between April 9, 2021, and April 5, 2022, recruited patients less than 12 years of age with congenital heart disease (CHD) from the general pediatric wards at Kenyatta National Hospital (KNH), Nairobi. CHD diagnosis was confirmed via echocardiogram by a pediatric cardiologist at KNH. Patients were to be followed up for one year. Exclusion criteria included parental refusal to consent or inaccessibility by mobile phone. Follow-up involved contacting parents/guardians one-year post-recruitment for additional data, focusing on the age at which CHD was suspected and confirmed, age at intervention, and follow-up status. Diagnosis ideally occurred on or before the first day of life, adhering to evidence-based standards (Park, 2014).

Patients were classified as "followed-up" if they had a pending pediatric cardiology outpatient (PCOPC) appointment at the time of the follow-up call. Follow-up was deemed complete if patients received appropriately scheduled PCOPC appointments, attended each appointment, and had a pending PCOPC appointment at the one-year follow-up call. PCOPC appointments were considered "appropriately given" if scheduled at discharge and after each subsequent visit. Complete follow-up also included patients who passed away before attending a PCOPC appointment. Patients were labeled as "lost-to-follow-up" if their parent/guardian couldn't be reached one year post-recruitment or didn't have a pending PCOPC visit during the one-year follow-up call. The Chi-square test was employed to compare current findings with those from 16 years ago (Awori et al., 2007).

RESULTS

Out of a total of 49 patients recruited, only 2

Table 2. Types of CHD.

Lesion	No. (%)
Complete atrioventricular canal defect	9(18.40)
Patent ductus arteriosus	6(12.20)
Ventricular septal defect	6(12.20)
Tetralogy of Fallot	6(12.20)
Double outlet right ventricle	4(8.20)
Coarctation of the aorta	3(6.10)
Transposition of great arteries	3(6.10)
Atrial septal defect	2(4.10)
Tricuspid atresia	2(4.10)
Truncus arteriosus	1(2.00)
Aorto-pulmonary window	1(2.00)
Absent right pulmonary artery	1(2.00)
Double inlet left ventricle	1(2.00)
Pulmonary stenosis	1(2.00)
Pulmonary atresia with intact ventricular septum	1(2.00)
Total anomalous pulmonary venous connection	1(2.00)
Hypoplastic left heart syndrome	1(2.00)
TOTAL	49(99.6)

Table 3. MP performance.

Parameter	Value
Age CHD suspected (mean)	35.3(SD+/-91.1) weeks
Age CHD confirmed (mean)	43.4(SD+/- 97) weeks
Healthcare facility where CHD confirmed	Public: 84%; private:16%
Locality where CHD confirmed	Nairobi: 86%; other: 14%
Intervention status	Had intervention: 34%, did not have: 66%
Age at intervention (mean)	47.5(SD +/- 51) weeks
Number of patients who died before intervention	23 (49%)
Kenya Healthcare facility where intention took place	Public: 100%; private: 0%
Locality where surgery took place	Abroad: 13%; Kenya: 87%
Types of surgery	Corrective: 60%; palliative: 40%
Operative mortality	Corrective surgery: 33%; palliative surgery: 50%
Patients being followed-up	94%
Status of patients at 1-year follow-up phone call.	Alive: 30%, dead: 70%

patients could not be reached during the 1-year follow-up phone call. Data was complete for 47 patients (95.6%). The types of congenital heart disease (CHD) and performance metrics of the Management Pathway (MP) are presented in Tables 2 and 3 respectively. The median ages when CHD was first suspected and diagnosed were 9 weeks (IQR 25) and 13 weeks (IQR 24) respectively, with a median age at intervention of 25.5 weeks (IQR 54.5). Among the patients who received intervention, 15 out of 16 (94%) underwent surgery, while one patient underwent balloon valvotomy. None of the patients were diagnosed or received intervention in accordance with the evidence-based protocol (Park, 2014), indicating a lack of optimal care (OC) in the study cohort.

A follow-up rate of 96% was achieved, with none of the patients receiving OC. Among the 14 patients still alive at the 1-year mark post-recruitment, 10 (71%) had complete follow-up. The suspicion and confirmation of CHD occurred primarily at a public health facility. Notably, only 35% of patients underwent surgery, with a concerning operative mortality rate of 39% among those operated on in Kenya (margin of error +/-27%; 95% Confidence Interval). Alarmingly, over 70% of all patients had passed away by the 1-year mark post-recruitment, with 49%

deaths occurring before any intervention could be administered.

DISCUSSION

The mean ages at which congenital heart disease (CHD) was first suspected and diagnosed were 35.3 and 43.4 weeks respectively, with the mean age at intervention being 47.5 weeks; these figures represent approximately half of what they were 16 years ago (Awori et al., 2007). The proportion of patients diagnosed at Kenyatta National Hospital (KNH) was significantly lower (76 vs. 94%) compared to 16 years ago (Awori et al., 2007). Furthermore, 96% of patients were followed up, a significant improvement from the 46% follow-up rate recorded 16 years ago (Awori et al., 2007). These findings suggest a notable improvement in at least two phases of the Management Pathway (MP), indicating the potential for patients with critical CHD to now receive timely intervention.

However, despite evidence of improvement in the MP, none of the patients experienced optimal care (OC), and the proportion of patients receiving surgery did not significantly differ from what it was 16 years ago (35 vs. 50%). Studies conducted in Pakistan and Laos examining two aspects of the MP (diagnosis and treatment) revealed that no patients received care in accordance with evidence-based protocols (Hwang et al., 2017; Mostafa et al., 2021). Conversely, a study from India demonstrated that the same two phases of the MP could be delivered in accordance with evidence-based protocols (Reddy et al., 2015). These collective findings from Asia suggest that while challenges persist in delivering the CHD MP according to evidence-based protocols, it is achievable even in resource-limited environments (Reddy et al., 2015).

Timely diagnosis/intervention and robust follow-up are essential features of an optimal Congenital Heart Disease Management Pathway (CHD MP). The Gross Domestic Product (GDP) per capita serves as an indicator of a country's wealth, and generally, healthcare quality improves as both GDP per capita and healthcare spending increase (Our World in Data, 2023). Kenya's GDP per Capita, adjusted by purchasing power parity rates, rose from USD 2,481 in 2007 to USD 5,764 in 2022 (World Bank, 2023), which likely contributed to the enhancement of the MP's quality. Additionally, as a country becomes wealthier, the number of urban centers increases, leading to skilled personnel migrating to and settling in these areas, potentially improving the MP's quality. The reduction in the proportion of patients diagnosed at Kenyatta National Hospital (KNH) could be a manifestation of this trend.

Furthermore, Kenya's adoption of a decentralized system of government in 2010 facilitates better resource utilization across the country, which could also contribute to the MP's improvement (Kimenyi, 2013). Despite the achievement of good surgical results in Low- and Middle-Income Countries (LMICs), the operative mortality of patients operated on in Kenya remains high, possibly around 12%, which is double what it should be. This might be due to the low surgical load at KNH and delayed surgeries, leading to patients presenting with greater morbidity at the time of surgery.

There's on-going concern regarding the cost justification of CHD surgery in LMICs, with arguments favoring public health measures over expensive surgeries. However, the Disability Adjusted Life Year (DALY) was developed to facilitate rational cost-benefit analyses in healthcare spending, revealing that pediatric cardiac surgery (PCS) is as cost-effective as some common public health measures (Grimes et al., 2014). Some even propose listing PCS as an essential pediatric surgical procedure (Saxton et al., 2016). The introduction of the DALY is significant as it enables LMIC governments to justify resource allocation toward developing PCS services.

The study has some limitations: the sample size is small and may not be representative of the true spectrum of congenital heart disease (CHD) encountered in Kenya. This potential bias could result in an over-representation of more complex forms of CHD, potentially leading to an increased mortality rate during follow-up. Additionally, due to the one-year follow-up period, the quality of long-term follow-up remains unknown. Nonetheless, these limitations have not significantly impacted the ability to detect improvements in the Management Pathway (MP) or identify areas in need of improvement.

Conclusion

We observed that at least two phases of the Management Pathway (MP) may have improved over the past 16 years. Specifically, the age at diagnosis has been halved, suggesting that children with Critical Congenital Heart Disease (CCHD) may now receive intervention. The follow-up phase has also seen improvement, with a significant increase in the proportion of patients being followed up. However, despite these improvements, approximately half of the patients die before receiving surgery, and for those who do undergo surgery, the operative mortality remains higher than expected. Based on Disability Adjusted Life Years (DALYs) averted by Pediatric Cardiac Surgery (PCS), Low- and Middle-Income Countries (LMICs) are justified in allocating funds to improve the CHD MP, with a priority on the treatment phase (especially surgery).

CONFLICT OF INTERESTS

The authors have not declared any conflict of interests.

REFERENCES

Awori MN, Ogendo SW, Gitome SW, Ong'uti SK, Obonyo NG (2007).

Management pathway for congenital heart disease at Kenyatta National Hospital, Nairobi. East Africa Medical Journal 84(7):312-317.
Fleming S (2022). The World Bank's 2020 country classification explained. Available at https://www.weforum.org/agenda/2020/08/world-bank-2020-classifications-low-high-income-countries. Accessed December 12, 2022.
Gan HL, Zhang JQ, Zhou QW, Feng L, Chen F, Yang Y (2014). Patients with congenital systemic-to-pulmonary shunts and increased pulmonary vascular resistance: what predicts postoperative survival? PLoS One 9(1):e83976.
Glidewell J, Olney RS, Hinton C, Pawelski J, Sontag M, Wood T, Kucik JE, Daskalov R, Hudson J (2015). State Legislation, Regulations, and Hospital Guidelines for Newborn Screening for Critical Congenital Heart Defects - United States, 2011–2014. Centers for Disease Control and Prevention (CDC). Morbidity and Mortality Weekly Report 64(23):625-630.
Grimes CE, Henry JA, Maraka J, Mkandawire NC, Cotton M (2014). Cost-effectiveness of surgery in low- and middle-income countries: a systematic review. World Journal of Surgery 38(1):252-263.
Hwang IC, Sisavanh M, Billamay S, Phangmanixay S, Oudavong B, Kang J, Kwon BS, Kim GB, Bae EJ, Noh CI, Choi JY (2017). Congenital heart disease at Laos Children's Hospital: Two year experience. Pediatrics International: Official Journal of the Japan Pediatric Society 59(3):271-279.
Kimenyi MS (2023). Devolution and resource sharing in Kenya. Available at https://www.brookings.edu/articles/devolution-and-resource-sharing-in-kenya/; accessed 14th August 2023.
Liu Y, Chen S, Zühlke L, Black GC, Choy MK, Li N, Keavney BD (2019). Global birth prevalence of congenital heart defects 1970-2017: updated systematic review and meta-analysis of 260 studies. International Journal of Epidemiology 48(2):455-463.
Mostafa H, Rashed M, Azzo M, Tabbakh A, El Sedawi O, Hussein HB, Khalil A, Bulbul Z, Bitar F, El Rassi I, Arabi M (2021). Congenital Heart Disease in Syrian Refugee Children: The Experience at a Tertiary Care Center in a Developing Country. Pediatric Cardiology 42(5):1010-1017.
Our world in data (2023). Healthcare spending versus Gross domestic product. Available at https://ourworldindata.org/grapher/healthcare-expenditure-vs-gdp Accessed 7th August 2023.
Park MK (2014). Pediatric Cardiology for Practitioners E-Book: Expert Consult-Online and Print. Elsevier Health Sciences.
Reddy NS, Kappanayil M, Balachandran R, Jenkins KJ, Sudhakar A, Sunil GS, Raj RB, Kumar RK (2015). Preoperative Determinants of Outcomes of Infant Heart Surgery in a Limited-Resource Setting. Seminars in Thoracic and Cardiovascular Surgery 27(3):331-338.
Samanek M (1992). Children with congenital heart disease: probability of natural survival. Pediatric Cardiology 13(3):152-158.
Saxton AT, Poenaru D, Ozgediz D, Ameh EA, Farmer D, Smith ER et al. (2016). Economic Analysis of Children's Surgical Care in Low- and Middle-Income Countries: A Systematic Review and Analysis. PLoS One 11(10):e0165480.
World Bank (2023). World development indicators. Available at Worldbank; https://data.worldbank.org/indicator/NY.GDP.PCAP.PP.CD?locations=KE; accessed 7th August 2023.