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Characteristics and outcomes in paediatric patients presenting with congenital colorectal diseases in sub-Saharan Africa

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Abstract

In sub-Saharan Africa, there is a high burden of paediatric surgical conditions and a paucity of data regarding outcomes of congenital colorectal anomalies. A retrospective, descriptive analysis utilizing the Kamuzu Central Hospital (Lilongwe, Malawi) paediatric acute care surgery database (age $\leq$ 18 years) over 44 months was performed. Of the 133 children presenting, 82 had Hirschsprung disease (HD) (2.4 $\pm$ 2.7 years) and 51 had anorectal malformations (ARM) (1.8 $\pm$ 2.4 years). Of the latter, 51.0% underwent surgery, mainly exploratory laparotomy (n = 15, 57.7%) and posterior sagittal anorectoplasty (n = 7, 26.9%). Of those with HD, 50.0% underwent operative intervention (77.3% boys), including exploratory laparotomy (n = 17, 41.5%) and definitive pull-through (n = 8, 19.5%). A dearth of expert paediatric surgeons and limited exposure to paediatric conditions in general surgeons limits definitive surgery. An emphasis on paediatric surgical training and improvement of referral networks for definitive therapy will improve patient outcomes.

Keywords

Anorectal malformation, Hirschsprung disease, sub-Saharan Africa, paediatric surgery, global surgery

Introduction

Congenital anomalies are a major cause of paediatric and neonatal mortality worldwide with an estimated incidence of 12 per 1000 live births globally.[1] Low- and middle-income countries (LMIC) bear the burden of congenital anomalies and resultant mortality, with an estimated 90% occurring in these countries.[2] Unfortunately, this is a gross underestimation owing to the paucity of quality data on congenital anomalies. The disparity in burden and outcomes data of congenital anomalies between high- and low-income countries is attributable to the establishment of epidemiological surveillance networks of congenital diseases in high-income countries (HIC). The reduction of mortality in HICs is also related to an improvement in medical and surgical management (including prenatal and early postnatal diagnosis) of neonates with an appropriate paediatric surgical workforce and expertise present.

Congenital colorectal diseases, including anorectal malformation (ARM) and Hirschsprung disease (HD), are the two most commonly managed cases by paediatric surgeons. The global incidence of ARM and HD are estimated to affect one in 4000–5000 live births and one in 5000 live births, respectively.[3–6] Congenital colorectal anomalies compose a significant surgical burden in LMICs, accounting for nearly 50% of cases of paediatric intestinal obstruction in Malawi (18% ARM, 29% HD) and 30% in Nigeria (ARM 16%, HD 14%).[7,8] Furthermore, delayed diagnosis is common.[9] A significant proportion of patients present with late signs and symptoms of intestinal obstruction

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because of failure of diagnosis at birth, principally because most births occur outside organized healthcare.

Studies from HICs show an apparent lower incidence of ARMs in the black population compared to Caucasians.[10] In contrast, studies by Louw et al. in South Africa showed an overall incidence of 5.5 cases of ARM per 10,000 live births, with 4.4 per 10,000 in Black Africans, which is higher than the reported global incidence.[11,12]

We aimed to characterize congenital colorectal anomalies in a Malawian tertiary care centre and described both operative management and patient outcomes.

Methods

We conducted a retrospective descriptive analysis of a hospital-based paediatric acute care surgery surveillance database between February 2012 and October 2015 at Kamuzu Central Hospital (KCH) in Lilongwe, Malawi. All paediatric surgery patients aged <18 years, who presented to the emergency department with a surgical diagnosis, admitted to the paediatric ward with a surgical diagnosis, or underwent surgical consultation, were recorded in the database. Children with the congenital colorectal anomalies, HD and ARM, were included in the analysis.

KCH is a 600-bed hospital and tertiary referral centre for central Malawi, with a catchment area of about six million people. KCH contains dedicated children's wards including a surgical ward, a paediatric high-dependency unit and intermediate care ward. During the study period, there were four full-time general surgeons, no paediatric surgeons, three full-time orthopaedic surgeons, multiple clinical officers and an 11-resident general surgery residency-training programme. The operating theatre at KCH has four operating rooms with one anaesthesiologist and six clinical officer anaesthetists. Diagnostic radiology capacity includes plain and contrast radiological studies, as well as abdominal ultrasound. Computed tomography and magnetic resonance imaging were not available. There was no pathologist on staff at KCH during the time of the study.

Demographic data, including sex and age, were collected in this study. Clinical variables collected included: type of admission; diagnosis; time to operative intervention; operative intervention performed; hospital length of stay; and discharge disposition. The KCH paediatric surgery database is kept prospectively. Initial admission diagnosis is entered; as information is gathered during the clinical course, the database is updated.

The National Health Science and Research Council of Malawi and the institutional review board of the University of North Carolina at Chapel Hill approved this study.

Results

During the 20-month study period, 1680 paediatric surgical admissions and consultations were seen at KCH. Of these, 133 paediatric patients with congenital anorectal disease presented. HD and ARM were diagnosed in 82 (61.7%) and 51 (38.3%) patients, respectively. HD was found predominantly in boys (n = 58, 70.7%) with a mean age of 2.4 ± 2.7 years. ARM was found primarily in girls (n = 30, 58.8%) with a mean age of 1.8 ± 2.4 years (Table 1).

Of those with ARM, 26 (51.0%) underwent operative intervention, with an average age of 1.5 ± 2.5 years (median = 0.8 years) at time of operation and average time from presentation to operative intervention of 12.7 ± 15.7 days. Procedures performed for ARM were exploratory laparotomy with diverting colostomy in 15 patients (57.7%), followed by posterior sagittal anorectoplasty (PSARP) in seven patients (26.9%) and anal dilation in three patients (11.5%). Four of these specific interventions were performed as emergencies; all underwent an emergency exploratory laparotomy with colostomy. Fistula dilation and irrigations were the standard of non-operative management for ARM. Length of stay for operated patients was 7.7 ± 69.7 days (median = 12 days) compared to 8.8 ± 11.6 days (median = 4 days) for patients treated conservatively.

Two children with ARM died during their admission, while the remaining 24 were discharged home. All but one of the non-operative ARM cases were discharged home (n = 23); one patient absconded.

An equal number (50%, n = 41) with HD underwent operative and non-operative management. Boys composed 75.6% (n = 31) with a mean age of 2.7 ± 3.1 years (median = 1.2 years) at presentation. The average time from presentation to operative intervention was 11.7 ± 13.8 days, being exploratory laparotomy in 17 patients (41.5%) followed by rectal biopsy in 14 patients (34.1%) and a definitive pull-through operation in eight patients (19.5%). Of the four with HD who had emergency procedures, three underwent exploratory laparotomy with colostomy and one had a definitive pull-through operation.

The mean age at presentation in the non-operative cohort was 2.2 ± 2.3 years (median = 1.4 years).

All these children were given rectal enemas. The length of hospital stay of operated HD patients was, not surprisingly, significantly longer (17.9 ± 12.1 days) compared with those who did not undergo surgery (9.7 ± 7.9 days). One child died following surgery, but all others survived. Thus, only 9.8% and 13.7% having HD and ARM, respectively, underwent definitive surgery.

During the course of our study, we could not obtain proper follow-up data, as most children did not return, mainly for logistical reasons.

Table 1. Patient characteristics of patient in the KCH paediatric acute care surgery surveillance database diagnosed with HD and ARM.

	Anorectal malformation (n = 51)			Hirschsprung disease (n = 82)		
	Surgical management (n = 26)	Non-surgical management (n = 25)	All patients (n = 51)	Surgical management (n = 41)	Non-surgical management (n = 41)	All patients (n = 82)
Patient age (years) (mean ± SD)	1.5 ± 2.5	1.9 ± 2.4	1.8 ± 2.4	2.7 ± 3.1	2.2 ± 2.3	4.2 ± 4.3
Gender (n (%))						
Male	11 (42.3)	10 (40.0)	21 (41.2)	31 (75.6)	27 (65.9)	58 (70.7)
Female	15 (57.7)	15 (60.0)	30 (58.8)	10 (24.4)	14 (34.1)	24 (29.2)
Missing	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)
Surgery type (n (%))						
Exploratory laparotomy	15 (57.7)	–	15 (29.4)	17 (41.5)	–	17 (20.7)
PSARP	7 (26.9)	–	7 (13.7)	0 (0.0)	–	0 (0.0)
Pull-through	0 (0.0)	–	0 (0.0)	8 (19.5)	–	8 (9.8)
Dilation	3 (11.5)	–	3 (5.9)	0 (0.0)	–	0 (0.0)
Biopsy	0 (0.0)	–	0 (0.0)	14 (34.1)	–	14 (17.1)
Stoma closure	1 (3.8)	–	1 (2.0)	2 (4.9)	–	2 (2.4)
Disposition (n (%))						
Discharged	24 (92.3)	23 (92.0)	47 (92.2)	40 (97.6)	36 (87.8)	76 (92.7)
Death	2 (7.7)	0 (0.0)	2 (3.9)	1 (2.4)	0 (0.0)	1 (1.2)
Abscond	0 (0.0)	1 (4.0)	1 (2.0)	0 (0.0)	4 (9.8)	4 (4.8)
Missing	0 (0.0)	1 (4.0)	1 (2.0)	0 (0.0)	1 (2.4)	1 (1.2)

PSARP, posterior sagittal anorectoplasty.

Discussion

Approximately 50% of all children presenting with congenital colorectal malformation are managed without any operative interventions, let alone diagnostic or therapeutic intervention. The majority of operative intervention is staged and not definitive. There are significant challenges to paediatric surgical management in a resource-poor setting.

First, because most children are born outside a structured healthcare system and in the absence of a physician or indeed a trained nurse or midwife, there is failure of early recognition of ARM leading to delayed presentation or mortality before receiving any medical care.[13–15] Failure to pass meconium as a diagnostic indicator of congenital colorectal anomalies in the first 24 h of life is only helpful if the mother and neonate are admitted for 24 h. Hence, most patients present in distress days after birth with signs and symptoms of intestinal obstruction. There is also an increased incidence of perforation, megarectum and dilated colon which requires stoma formation and negates the option for a single-staged definitive procedure.[14] Ostomies performed by non-paediatric trained general surgeons may increase the risk of downstream complications.[16] Patients with ARM should have an endostomy and mucus fistula placed at the junction between the descending and sigmoid colon to provide complete diversion, decrease the risk of genitourinary infection and complications, and to improve results definitive operations. Delayed presentation was associated with a 19–35% morbidity in this patient population and a mortality of 4–10%.[14,17]

Second, there is a dearth of diagnostic adjuncts available in LMICs, as well as lack of specialist paediatric anaesthesiology, radiology, ultrasonography and pathology services.[18] In a survey performed in 2007, only 13% of Ugandan anaesthetists believed they had the appropriate tools to take care of children aged <5 years.[19,20] In HD, a pathologist is required to make the diagnosis and identify the level of aganglionosis.

Furthermore, there is a significant surgical workforce shortage. Trained general surgeons have varying comfort levels performing paediatric procedures. Thus, very few PSARPs and pull-through procedures are performed. This is not solely a Malawian problem.[18]

As about half of the children with HD and ARM did not receive any surgical management at all, it is likely the majority of these children would have suffered fatal intestinal obstruction or continued with chronic persistence of partial obstructive symptoms. Some may have found intervention elsewhere, but at the time of the study the nearest paediatric surgeon was based in Blantyre, a 350-km distance.

Conclusion

This study of the characteristics of congenital colorectal anomalies in a sub-Saharan Africa tertiary care centre indicates the need for a focused educational curriculum to diagnose congenital colorectal anomalies directed towards medical providers, community health workers and nurse midwives. This may limit the delay of patient diagnosis and encourage early surgical referral. General surgeons are often the first surgical contact. Local general surgical training programmes should incorporate paediatric surgical skills training with emphasis on staged operative interventions and optimal referral networks for definitive surgical management to the closest paediatric surgical centre. Expanding referral networks between hospitals without paediatric surgery and paediatric surgical centres is imperative. Alternatively, partnerships with existing paediatric surgical specialists to visit tertiary centres without a full-time paediatric surgeon may help mitigate patient referral for definitive surgical care. As the training of surgical personnel in paediatric surgery in sub-Saharan Africa matures, increased availability of adjunct specialties, such as paediatric anaesthesia and critical care may attenuate morbidity and mortality in the paediatric surgical population.

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