

Original Article

© 2024 Abrahams et al.

Submitted: 21-08-2023 **Accepted:** 29-11-2023

License: This work is licensed under a <u>Creative Commons Attribution 4.0</u> <u>International License</u>.

DOI: https://doi.org/10.47338/jns.v13.1250

The outcomes of specific surgically correctable congenital gastro-intestinal malformations at a tertiary level neonatal intensive care unit in South Africa

I Abrahams,1*C de Vos,2 L Van Wyk1

- 1. Division of Neonatology, Dept. Paediatrics & Child Health, Stellenbosch University, Cape Town, South Africa
- 2. Division of Paediatric Surgery, Stellenbosch University, Cape Town, South Africa

Correspondence*: Ilhaam Abrahams, MBChB, MMed (paed) Department of Paediatrics and Child Health, Faculty of Medicine and Health Sciences, Stellenbosch University, PO Box 241, Cape Town 8000, South Africa. E-mail: ilhaama@sun.ac.za

KEYWORDS

Congenital gastro-intestinal malformations, Surgery, Low-middle income, Neonates, Outcomes

ABSTRACT

Background: The outcome of neonates with congenital surgically correctable gastro-intestinal (GIT) malformations is poorly described in low middle income countries.

Methods: A 5-year retrospective descriptive analysis of neonates admitted to a tertiary level neonatal intensive care unit (NICU), with congenital, surgically correctable GIT malformations, was performed. The primary outcome was the 30-day postoperative mortality as well as survival to 1 year. Secondary outcomes included patient demographics, clinical presentation as well as the burden of disease in our study population.

Results: Eighty-four neonates met study criteria. The mean gestational age was 35 weeks (SD 3.19) and birthweight 2518g (SD 789.3). The most common congenital malformations were intestinal atresia (39%) followed by omphaloceles (21%). Associated systemic malformations were common (39%). The majority of neonates (88%) underwent surgery at a median age of 2 days (IQR 1.5-5). Ventilation was required in 19% of neonates preoperatively and 65% postoperatively. Full feeds were achieved at a median age of 13 days (IQR 9-18) after surgery. The 30 day-postoperative survival rate was 97% with a survival to discharge or transfer of 86%. Data for one year survival was available for 80% of neonates with a one-year survival rate of 75%.

Conclusion: The 30-day postoperative survival was high in neonates with congenital, surgically correctable GIT malformations. Outcomes and burden on the healthcare system was dependent on the type of lesion. With early diagnosis and referral to a tertiary centre, good outcomes can be achieved. Neonates with congenital GIT malformations should have long term follow up to monitor growth and neurodevelopment as well as to address the high mortality post discharge.

INTRODUCTION

The Global Burden of Diseases Study in 2015 identified congenital malformations as the fifth leading cause of under-5 mortality, with 97% of these deaths occurring in low and middle-income countries (LMICs). [1] According to the World Health Organization (WHO), approximately 10% of neonatal deaths in Sub-Saharan Africa (SSA) and South Asia are attributed to congenital malformations, many of which are amenable to surgical intervention. [2] Moreover, significant long-term disability associated with congenital malformations is recognized as a global concern. [3]

Internationally, the incidence of various congenital gastrointestinal (GIT) surgical malformations varies from 1:2000 to 1:5000 live births. [4,5] A recent South African study reported an incidence of major

congenital malformations at 2.6/1000 live births, with the gastrointestinal tract being the most common site for congenital surgical malformations presenting in the neonatal period. [6]

Congenital GIT malformations may occur in isolation or be associated with other systemic malformations or syndromes. [9] Clinical presentations are often acute and life-threatening, underscoring the importance of prompt diagnosis and treatment. [8,10] Surgery has been identified as the most cost-effective health intervention, highlighting the necessity for a functional neonatal intensive care unit (NICU). [11,12]

Neonates with surgically correctable GIT malformations pose unique challenges, including delayed enteral feeding, prolonged admission, and a range of surgical complications such as abdominal compartment syndrome, short bowel syndrome, and

wound site infections. [9-16]. Emergency neonatal surgery in SSA is associated with mortality rates between 30–43%, with the highest mortality observed in neonates younger than 7 days old. [14] Specific malformations like esophageal atresia (EA), jejunoileal atresia, intestinal obstruction, congenital diaphragmatic hernia, and abdominal wall defects are linked to the highest mortality rates. [12] Causes of death are primarily attributed to sepsis and respiratory issues, with a smaller proportion linked to multiple severe malformations. [13,17]

In December 2020, the WHO released a toolkit aimed at enhancing the surveillance of global congenital anomalies to improve care, reduce the number of babies born with congenital anomalies, and enhance care for those affected. [15]

South Africa lacks a national congenital GIT malformation register, resulting in an unknown burden of disease and outcomes related to these malformations. This study seeks to determine the outcomes of congenital, surgically correctable GIT malformations in a resource-restricted public health hospital.

METHODS

A retrospective, descriptive study encompassing all neonates born at or transferred to the Neonatal Intensive Care Unit (NICU) at an academic hospital from January 1, 2014, to December 31, 2018, was conducted. This NICU operates within a resourcerestricted tertiary-level public health institution and serves as one of two neonatal surgical referral centers in the province. The study included neonates, except those with esophageal atresia (EA) and gastroschisis, who presented with congenital surgical gastrointestinal (GIT) malformations such duodenal atresia, intestinal atresia, midgut volvulus, intestinal malrotation, omphalocele, malformation, Hirschsprung's Disease, small left colon syndrome, vitelline intestinal duct, and congenital diaphragmatic hernia. Neonates born with EA and gastroschisis were excluded, as they have been independently reviewed by authors at our institution. [16–18]

Collected data included maternal and neonatal demographic information, as well as neonatal medical and surgical outcome data. The primary objectives were to describe both the 30-day postoperative and overall 1-year survival rates. The dataset incorporated patient demographics, clinical presentation, and postoperative morbidity factors such as duration of ventilation, need for inotropes, time to full feeds, duration of NICU stay and hospitalization, necessity and duration of total parenteral nutrition (TPN), postoperative sepsis, and the need for a secondary surgical procedure.

The study protocol received approval from the Stellenbosch University Health Research Ethics Committee (S21/02/021) and the National Health Research Department (WC_202202_002). For statistical analysis, Stata Statistical Software version 14 (College Station, TX: StataCorp LP) was employed. Continuous data were described using mean and standard deviation or median and interquartile range, as appropriate, while categorical data were expressed as numbers and percentages.

RESULTS

Demographics:

During the study period, 84 neonates with surgically correctable congenital gastrointestinal (GIT) malformations were admitted to the Neonatal Intensive Care Unit (NICU). The most prevalent malformations in the cohort included intestinal atresias (39%), omphalocele (21%), anorectal malformations (14%), and congenital diaphragmatic hernia (CDH) (12%).

Table 1: Demographics of study population

Variable	Total	Result				
	n=84					
Maternal demographics						
Maternal age (years), m	72*	28.6 (7.1)				
Gravidity, median (IQR	72	2 (1.0-3.5)				
Parity, median (IQR)	72	2 (1.0-3.0)				
Booked, n (%)	82	79 (96)				
Mode of delivery, n	NVD		47 (56)			
	C/S	84	36 (43)			
	Assisted delivery		1 (1)			
Neonatal demographics						
Gestation in weeks, me	83	35.7 (3.19)				
Preterm, n (%)	83	44 (53)				
Birth weight, grams, m	84	2518.8 (789.3)				
Inborn, n (%)	83	42 (51)				
Male, n (%)	84	40 (48)				
RVD exposed, n (%)	84	10 (12)				

Maternal and neonatal demographic details are outlined in Table 1. The majority of mothers received antenatal care (>1 visit). Documentation of maternal illnesses and substance use was inadequate. Approximately 68% of mothers underwent at least one antenatal ultrasound, though not all were detailed scans. A significant proportion of babies were preterm (<37 weeks gestational age).

Clinical Presentation:

The clinical presentation, summarized in Table 2, revealed that a third of babies were diagnosed through antenatal ultrasound, with omphalocele or CDH being the most common diagnoses. Bilious

vomiting and delayed passage of stool were frequent clinical symptoms. Hemodynamic compromise was diagnosed in 11% of neonates, and 50% of CDH cases required treatment for hemodynamic compromise.

Thirteen neonates (16%) were treated for presumed sepsis at admission, predominantly those diagnosed with intestinal atresia (25%).

Table 2: Clinical presentation

Variable	Total	IA	Omphalocoele	CDH	Malrotation and volvulus	ARM	Other*
N (%)	84	33 (39)	18 (21)	10 (12)	5 (6)	12(14)	6 (7)
Antenatal U/S, n (%)	56 (68)	21(64)	15(83)	6 (60)	1(20)	8(67)	5(83)
Antenatal diagnosis, n (%)	30 (36)	11 (33)	12 (67)	6 (60)	0	1 (8)	0
Defect seen at birth, N (%)	25 (30)		18 (100)	0	0	6 (50)	1 (17)
Bile-stained vomiting n (%)	34 (43)	24 (73)	0	0	5 (100)	2(9)	1 (17)
Abdominal distension n/N (%)	28/78 (36)	14 (42)	0	0	4 (80)	5 (42)	5 (83)
Delayed passage of stool, n (%)	25/54 (46)	11 (33)	0	0	1 (20)	10 (83)	3 (50)
Age at NICU presentation, days, median (IQR)	2 (1-4)	2 (1-4)	1 (0-4)	0 (0-5)	3 (3-4)	2 (1-3)	9 (5-21)
Ventilation pre-op, n/N (%)	16/81 (20)	3/32 (9)	2/17 (12)	9 (90)	1/4 (25)	0	1 (20)
INR, mean (SD)	1.3 (0.5)	1.4 (0.7)	1.3 (0.3)	1.4 (0.4)	1.2 (0.2)	1.4 (0.3)	1.1 (0.1)
pH on admission, mean (SD)	7.4 (0.1)	7.4 (0.1)	7.3 (0.7)	7.2 (0.2)	7.4 (0.9)	7.4 (0.7)	7.4 (0.9)
Admission lactate, mmol/l, mean (SD)	3.1 (2.1)	3.1 (1.7)	2.7 (1.6)	3.0 (2.1)	2.9 (1.6)	3.2 (1.9)	3.7 (5.2)

ARM= anorectal malformation; CHD= congenital diaphragmatic hernia; IA= intestinal atresia; INR= international normalized ratio; IQR= interquartile range; NICU=neonatal intensive care unit; SD= standard deviation; U/S= ultrasound * other includes 2 cases of Hirschsprung's Disease, 1 case each of small left colon syndrome, microcolon, pyloric stenosis and vitelline intestinal duct.

Associated congenital malformations were common, with 38% having cardiac anomalies, 28% renal anomalies, and 13% limb anomalies, including 16% defined as VACTERL anomalies. Genetic testing was conducted in 45% of neonates, with Trisomy 21 being the most common diagnosis.

Surgery and Post-operative Course:

Eighty-eight percent of neonates underwent surgery (Fig. 1). The median time to the first feed was just over 1 week, with full feeds achieved at a median of 13 days postoperatively. Time to feed varied between lesions, with neonates having omphalocele achieving full feeds earliest at a median time of 7 days postoperatively (Table 3).

Morbidity and Mortality:

Postoperative morbidity and mortality are summarized in Table 3. The overall survival for the entire study population (including those who did not undergo surgery) was 86%, and the 30-day postoperative survival was 97%. Two neonates died post-operatively, one due to sepsis and the other due to total bowel necrosis diagnosed at surgery.

The length of NICU admission was a median of 9 days, and the length of hospital stay was a median of

19 days. NICU admission duration and hospitalization length differed between types of lesions.

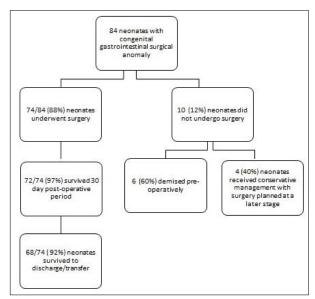


Figure 1: Study population

Postoperative sepsis was present in 45% of all neonates, with gram-negative organisms being the most common. Nearly 10% of neonates required at least one readmission to the NICU for various reasons.

Twelve percent of neonates underwent a second surgical procedure while still in the neonatal service.

Table 3: Surgery and post-operative outcomes

Variable	Total	IA	Omphalocoele	CDH	Malrotation and volvulus	ARM	Other
n	84	33	18	10	5	12	6
Surgery n (%)	74(89)	33(100)	15/18 (83.3)	4 (40)	5 (100)	12 (100)	5 (83)
Day of life surgery, median (IQR)	2 (1.5-5)	3 (2-5)	1 (1-2)	7 (4.5-17)	3 (3-4)	2 (1.5-3)	10 (5-21)
Ventilated post-op, n/N (%)	48/74 (65)	23/32 (72)	6/15 (40)	4 (100)	2 (40)	8 (67)	5 (100)
Duration of post -op ventilation in days, median (IQR)	2 (1-4)	2 (1-4)	1 (1-4)	17 (8-26.5)	1.5 (1-2)	2 (1.5-3)	3 (2-4)
Inotropes post -op, n/N (%)	4/74 (5)	1/32 (31)	0	1 (25)	0	2 (17)	0
Duration of inotropic support in days, median (IQR)	2 (1.5-9.5)	2		10 (10-10)		1.5 (1-2)	
CVP , n (%)	63/83 (76)	32 (100)	8/(44)	(90)	2 (40)	7 (58)	5 (83)
Duration of CVP in days, median (IQR)	8.5 (4-12.5)	9 (6-13)	5 (1.5-14)	2.5 (1-10)	6.5 (5-8)	8 (4-12)	11 (9-19)
TPN , n/N (%)	54 (65)	31/32 (100)	5 (28)	5 (50)	2 (40)	6 (50)	5 (83)
Duration of TPN in days, median (IQR)	8.5 (5-13)	9 (5-12)	11 (6-15)	8 (7-14)	5.5 (7-4)	8.5 (5-11)	10 (8-19)
Post-op sepsis, n/N (%)	33/73 (45)	13/32 (41)	8/15 (53)	3/4 (75)	2 (40)	5 (42)	2/5 (40)
Day to first feed post- surgery, median (IQR)	8 (5-10)	12 (8-15)	5 (3-14)	2.5 (0-18)	8 (6-9)	8 (1-14)	9 (6-11)
Day to full feeds post- surgery, median (IQR)	13 (9-18)	15 (11-18)	7 (5-14)	28.5 (22.5-33)	12 (11-13)	12 (5-16)	19.5 (15-27)
Secondary procedure, n/N (%)	9/74 (12)	3 (9)	3 (16)	0	1 (20)	2 (16)	0
Duration of initial NICU stay, days, median (IQR)	9 (5-18)	12 (8-18)	4 (4-8)	9.5 (1-29)	11 (10-12)	8.5 (5.5-15.5)	23.5 (11-27)
Duration of hospitalization , days, median (IQR)	19 (12-31)	19 (14-28)	10 (7-23)	39.5 (31.5-91)	16 (12-16)	24 (11-41)	28 (20-37)
30-day post-operative survival, n/N (%)	72/74 (97)	32/33 (97)	15/15 (100)	4/4 (100)	5/5 (100)	11/12 (92)	5/5 (100)
Survival to discharge/transfer**, n/N (%)	72/84 (86)	31/33 (94)	16/18 (89)	3/10 (30)	5/5 (100)	11/12 (92)	6/6 (100)
Age at death, days, median (IQR)	14.5 (8.5-32)	15 (12-16)	39.5 (18.5-50)	6.5 (1-22)		20.5 (11-30)	20

ARM= anorectal malformation; CHD= congenital diaphragmatic hernia; CVP= central venous pressure; IA= intestinal atresia; IQR= inter quartile range; NICU=neonatal intensive care unit; SD= standard deviation; TPN= total parenteral nutrition* Other includes 2 cases each of Hirschsprung's Disease and 1 case each of small left colon syndrome, microcolon, pyloric stenosis and vitelline intestinal duct. ** Includes entire study population regardless if surgery occurred or not.

Table 4: Pre-existing TBH data on gastroschisis and oesophageal atresia

	Gastroschisis	Gastroschisis	Oesophageal atresia	
	Van Eck 2017 [17] N=31	Negobo 2023 [18] N=50	De Vos et al 2022 [16] N=53	
Time to extubation in days, mean (SD)	6.1 (±9)	4* (IQR 0-23)	7.6 (±9.2),	
Time to full feeds in days, mean (SD)	21 (±13)		17 (±12.6)	
Duration of NICU stay in days, mean (SD)	16.8 (±15.7)	23* (IQR 0-95)	16.6 (±14.0),	
Length of stay in days, mean (SD)	37.5 (±27)		31.5 (±29.5)	
Post-operative sepsis, n (%)	13 (46%)	27 (54%)		
30-day mortality, n (%)		7 (14%)	5 (9%)	
Survival to discharge/ transfer, n (%)	26 (84%)	39 (78%)		

SD = Standard deviation, NICU = neonatal intensive care unit, *median.

The 30-day post-operative mortality rate was 3%, with higher mortality rates observed in neonates born with CDH. At the end of the 30-day postoperative period, 28% of babies were still admitted to the hospital. Survival to discharge or step-down care transfer was 100% in certain groups of neonates, with malrotation,

volvulus, and mixed pathologies showing the highest survival rates, followed by those with intestinal atresia, anorectal malformations, and omphaloceles.

Data on 1-year survival was available for 80% of the study population, with 75% of them alive at 1 year of

age. Causes of mortalities for the deceased infants were not documented.

DISCUSSION

This retrospective audit looked at 30-day mortality and survival to one year in neonates with surgically correctable congenital GIT malformations at a single tertiary neonatal centre during 5 years. The overall survival to transfer or discharge was moderate with a high 30-day postoperative survival and a moderate 1-year survival. Our study did demonstrate significant sepsis rates.

An antenatal diagnosis was made in one-third of neonates in our study. Antenatal detection of congenital GIT malformations reported in the literature ranges from 9.4% to 85.7%. [19,20] While antenatal diagnosis has not been shown to be associated with improved survival, however it helped in prenatal counselling and planning the delivery. [21-23]

Associated malformations were present in more than one-third of our study population. Congenital GIT malformations may occur in isolation or be associated with other malformations or syndromes. [9] Trisomy 21 was identified in just over 10% of our study cohort that underwent genetic testing, and all cases were diagnosed with duodenal atresia. Trisomy 21 is known to be associated with duodenal atresia, duodenal web, and Hirschsprung's disease. [24-26] Similar to previously described, just over 80% of our study population with an anorectal malformation (ARM) had an associated malformation, some of which were associated with VACTERL. [27] Clinicians should be vigilant about the high incidence of associated anomalies in neonates with congenital surgical malformations, particularly those with ARM.

Surgery was performed at a median of 2 days in this study. The Global PaedSurg Collaboration study found the median age at presentation to be 3 hours (IQR 0-28) in high-income countries (HICs), 24 hours (IQR 3-9) in middle-income countries (MICs), and 72 hours (IQR 16-192) in low-income countries (LICs). [22] In this study, infants presented to the NICU at a median of 2 days of age (IQR 1-4), with the time of presentation depending on the lesion type. Delayed presentation may lead to the higher mortality rates seen in LMICs. [12] A Nigerian study found that less than 20% of neonates with ARM presented within 24 hours of birth. This delay in presentation may result in abdominal distention, which can result in respiratory compromise as well as fluid and electrolyte abnormalities. [24] In an Ethiopian review neonatal intestinal obstruction, presentation was found in 72% of cases. [25] Mortality rates as high as 74% have been reported prior to surgery, signifying the severe physiological

derangements associated with delayed presentation of these abnormalities. [14] Early diagnosis and presentation are essential to decrease the high mortality rates associated with congenital GIT abnormalities in LMICs.

Delays in establishing feeding in this cohort could be attributed to factors such as prematurity, NPO status in anticipation of extubation, and sepsis-related feed intolerance. Notably, one neonate with an anorectal malformation (ARM) and another with an omphalocele struggled to establish feeding and were subsequently diagnosed with duodenal atresia and ileal atresia, respectively. The Neonatal Intensive Care Unit (NICU) currently lacks a formal postoperative feeding guideline. While the implementation of postoperative feeding guidelines has demonstrated improved outcomes; collaborative care between the pediatric and neonatologist crucial surgeon is individualizing the feeding in this population. [28, 29]

Postoperative sepsis was present in nearly half of our study population, with the majority of cases attributed to gram-negative organisms. Postoperative sepsis has been identified as a major contributor to 30-day postoperative mortality, with a South African study reporting that 74% of its 30-day postoperative mortality was attributable to sepsis. [21, 26] Although our 30-day postoperative mortality due to sepsis (1/2; 50%) was not as high, there is concern about the presence of antimicrobial-resistant organisms. Allcause mortality due to infections from these organisms is high (12.5%-48%) in South African neonatal units. [30-32] Underlying surgical conditions and admission to a NICU are major risk factors for antimicrobial-resistant sepsis. [33] In many highincome countries, surgery-associated infections are predominantly caused by gram-positive organisms or fungi. [34] In our study, the minority of infections were gram-positive, and no fungi were cultured. Although previous studies have shown an increase in post-surgical mortality in premature neonates [35], this study did not specifically evaluate this interaction, despite half of the population being premature. Prioritizing early enteral feeding to reduce line days, adherence to infection prevention and control (IPC) strategies, and implementing antimicrobial stewardship (AMS) programs essential to reduce sepsis rates.

The median length of stay in this study was 3 weeks, with the shortest duration observed in cases of omphalocele and the longest in cases of CDH. This aligns with findings from a British study examining the hospital stay duration for neonates undergoing surgery, which reported the shortest stay for omphalocele at 12 days (SD 3-28) and the longest for CDH at 28 days (SD 7-99). [36] Both studies identified

significant variability in the length of hospital stay among different congenital lesions. This variability is attributed to factors such as the type and severity of the lesion, the presence of associated anomalies, and other considerations, including sepsis. Analyzing data on the length of hospital stay can be valuable for counseling parents both prenatally and postnatally, as well as for aiding in the planning and allocation of health resources and infrastructure.

Most of our neonates required resource intensive care like postoperative ventilation, CVC placement, TPN administration, and prolonged NICU-stay or hospitalisation. An Austrian study also documented resource intensive care required for congenital malformations and stressed on developing a national register to facilitate optimized care, rational resource allocation, and evaluation of outcomes. [37] South Africa does not have a formal congenital malformation register.

A recent global, multi-center, prospective study looking at the mortality from congenital GIT malformations in 74 countries found the 30-day mortality to be 39.8% in LICs; 20.4% in MICs, and 5.6% in HICs. [22] A South African study of both congenital and acquired abdominal surgical cases found the 30-day postoperative mortality was 11%. [21] Both studies included a broader range of diagnoses than the current study, had larger study numbers, and included both neonatal and pediatric patients. The global review study also excluded infants with CDH. [21,22] Our study's 30-day postoperative mortality (3%) was lower than that of the South African study as well as that of HICs in the prospective global review. [21,22]

Disease-specific mortality in the developing world is considerably higher than our reported mortality rates, with ARM at 5.4-25.4%, intestinal atresias at 34%, and omphaloceles at 45.7%. [38–42] Possible reasons for our lower mortality rates include the availability of a functional NICU, easy access to ventilation and TPN, an on-site pediatric surgical service, a relatively early median time to surgery, and well-established infrastructure such as roads and a functioning hospital referral and ambulance service.

The higher mortality rates in low- and middle-income countries (LMICs) may be attributed to delayed presentation, unavailability of NICU facilities, and a lack of trained staff. [12] Access to neonatal intensive care for seriously ill neonates and those requiring major surgery leads to more favorable outcomes. [14] While intensive care is available in most of South Africa and high-income countries (HICs), it is lacking in most of Sub-Saharan Africa. [43] Newborns managed where there are no NICU facilities available face challenges in appropriate monitoring and lack access to ventilators and other life-saving equipment.

[14] A review of West African pediatric surgical capacity revealed that neither NICU nor general ICU facilities were available in over 50% of the 37 hospitals reviewed, despite pediatric surgery taking place at those facilities. [44] A shortage of trained staff further compounds this problem. [14] Addressing these discrepancies in health care availability and infrastructure could potentially narrow the gap between mortality rates in low-income countries (LICs), middle-income countries (MICs), and HICs.

The deaths in our cases predominantly occurred in antenatally diagnosed CDH cases; other factors also contributed like right-sided lesions, liver herniation, unfavorable lung:head ratio, and pulmonary hypertension (not analyzed in this dataset). The absence of ECMO for neonatal CDH cases at our institution and potential hidden pre-operative mortality further complicates the picture. [45] The specific causes of CDH-related deaths at our unit require further investigation.

The 1-year mortality in our study was substantial (25%), aligning with another South African study's findings. [21] However, neither study assessed postdischarge outcomes, leaving uncertainty about the reasons for post-discharge mortality. The elevated 1year mortality in both South African studies underscores the need for thorough post-surgery assessments in neonates with congenital GIT abnormalities to identify contributing factors. Survivors in this population face an increased risk of neurodevelopmental impairment in cognitive, motor, and language domains, along with issues such as gut dysbiosis and growth impairment. [21,46] Notably, even minor procedures like pyloromyotomy have shown subtle neurodevelopmental differences at a 3year follow-up. [47] The impact of anesthesia and other interventions on neurodevelopmental outcomes remains unknown, emphasizing the importance of routine multidisciplinary follow-up, including neurodevelopmental screening, for this patient population. [47,48]

Limitations to this study included single centre study with small sample, and suboptimal documentation. Malformations with a high morbidity and mortality (gastroschisis and EA) were excluded from this study making interpretation of the low postoperative and 30-day mortality difficult. Patients who died before being admitted to the NICU were not accounted for. Long-term follow up of this study's population and review of cause of death for 1 year mortality was also not performed.

CONCLUSION

The overall survival of neonates with congenital GIT malformations was moderate, with a high 30-day survival rate among those undergoing surgery. However, this assessment is limited due to the

exclusion of cases involving EA and gastroschisis. The burden of disease, gauged by resource requirements, sepsis rates, and 1-year mortality in neonates post-surgery, is substantial. Establishing a national congenital surgical gastrointestinal registry is imperative. Long-term follow-up for these neonates is crucial to monitor growth, neurodevelopment, and address the elevated post-discharge mortality. Improving outcomes necessitates early recognition, timely referral to pediatric surgical centers, reduction in postoperative sepsis, and optimization of postoperative feeding.

REFERENCES

- Wang H, Bhutta ZA, Coates MM, Coggeshall M, Dandona L, Diallo K, et al. Global, regional, national, and selected subnational levels of stillbirths, neonatal, infant, and under-5 mortality, 1980–2015: a systematic analysis for the Global Burden of Disease Study 2015. Lancet [Internet]. 2016;388(10053):1725–74. Available from: http://www.sciencedirect.com/science/article/pii/S014067361 6315756
- Paul VK, Singh M. Regionalized perinatal care in developing countries. Semin Neonatol. 2004;9(2):117–24.
- Agarwal A, Rattan KN, Dhiman A, Rattan A. Spectrum of Congenital Anomalies among Surgical Patients at a Tertiary Care Centre over 4 Years. Int J Pediatr. 2017;2017:4174573-4.
- 4. Wright NJ. Management and outcomes of gastrointestinal congenital anomalies in low, middle and high income countries: protocol for a multicentre, international, prospective cohort study. Wright N, Leather A, Sevdalis N, Ade-Ajayi N, Ademuyiwa A, Ameh E, et al., editors. BMJ Open [Internet]. 2019;9(8). Available from: https://bmjopen.bmj.com/content/9/8/e030452 (Accessed 02/10/2019)
- Paoletti M, Raffler G, Gaffi MS, Antounians L, Lauriti G, Zani A. Prevalence and risk factors for congenital diaphragmatic hernia: A global view. J Pediatr Surg. 2020;55(11):2297–307.
- Mayer MMM, Velaphi SC. Incidence, types and outcomes of congenital anomalies in babies born at a public, tertiary hospital in South Africa. SAJCH. 2021;15(4):193-7.
- White R. Surgical emergencies. In: Roberts B, editor. Manual of clinical problems in Paediatrics. 5th ed. Lippincott Williams and Wilkins; 2000. p. 275–81.
- Morris G, Kennedy A, Cochran W. Small Bowel Congenital Anomalies: a Review and Update. Curr Gastroenterol Rep [Internet]. 2016 Mar;18(4):16. Available from: https://doi.org/10.1007/s11894-016-0490-4(Accessed 02/10/2019)
- Schindewolf E, Moldenhauer JS. Genetic counseling for fetal gastrointestinal anomalies. Curr Opin Obstet Gynecol. 2020;32(2):134–9.
- Desoky S, Kylat R, Udayasankar U, Gilbertson-Dahdal D. Managing neonatal bowel obstruction: clinical perspectives. Res reports Neonatol. 2018;8:19–32.
- Mock CN, Donkor P, Gawande A, Jamison DT, Kruk ME, Debas HT. Essential surgery: key messages from Disease Control Priorities, 3rd edition. Lancet. 2015;385(9983):2209–19.
- Ekenze SO, Ajuzieogu OV, Nwomeh BC. Challenges of management and outcome of neonatal surgery in Africa: a systematic review. Pediatr Surg Int [Internet]. 2016 Mar;32(3):291–9. Available from: https://doi.org/10.1007/s00383-016-3861-x
- 13. Klein MD. Chapter 75 Congenital Defects of the Abdominal Wall. In: Pediatric Surgery. 2012. p. 973–84.
- Ameh EA, Seyi-Olajide JO, Sholadoye TT. Neonatal surgical care: a review of the burden, progress and challenges in sub-Saharan Africa. Paediatr Int Child Health [Internet]. 2015;35(3):243-51. Available from:

Acknowledgements: Nil
Conflict of Interest: None.
Source of Support: Nil

Consent to Publication: Author(s) declared taking informed written consent for the publication of clinical photographs/material (if any used), from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient, however it cannot be guaranteed.

Author Contributions: Author(s) declared to fulfil authorship criteria as devised by ICMJE and approved the final version.

- http://www.tandfonline.com/doi/abs/10.1179/2046905515Y. 0000000033
- WHO. Birth defects surveillance: A manual for programme managers, 2nd Edition [Internet]. WHO, CDC IC for BD, editor. Geneva; 2020. 1–283 p. Available from: https://www.who.int/publications/i/item/9789240015395 (accessed 21/02/22)
- de Vos C, van Wyk L, Sidler D GP. The 30-day outcome of neonates operated for esophageal atresia. J Neonatal Surg. 2022:11:12.
- Van Eck A. A retrospective review of the outcomes of gastroschisis at a tertiary hospital in Cape Town. [Cape Town]: Master's Thesis, Stellenbosch University; 2017.
- Ngcobo Q, de Vos C. Risk factors for sepsis in neonates treated with gastroschisis at a tertiary hospital. Stellenbosch University: 2023.
- Kaur N, Pamnani S, Kaur B. Role of ultrasound in diagnosis of fetal congenital abdominal anomalies: One year prospective study. Int J Med Res Rev [Internet]. 2017;5(7):649–56. Available from: https://ijmrr.medresearch.in/index.php/ijmrr/article/view/89
 - https://ijmrr.medresearch.in/index.php/ijmrr/article/view/89 2
- Stefos TI, Plachouras NI, Sotiriadis A, Papadimitriou D, Almoussa N, Navrozoglou I, et al. Routine obstetrical ultrasound at 18-22 weeks: our experience on 7,236 fetuses. J Matern Fetal Med. 1999;82:64–9.
- 21. Siyotula T, Arnold M. An analysis of neonatal mortality following gastrointestinal and/or abdominal surgery in a tertiary hospital in South Africa. Pediatr Surg Int. 2022;38(5):721-9.
- Wright NJ, Leather AJM, Ade-Ajayi N, Sevdalis N, Davies J, Poenaru D, et al. Mortality from gastrointestinal congenital anomalies at 264 hospitals in 74 low-income, middle-income, and high-income countries: a multicentre, international, prospective cohort study. Lancet [Internet]. 2021;398(10297):325-39. Available from: https://www.sciencedirect.com/science/article/pii/S01406736 21007674
- Leather AJM, Ngwenya S, Jones B, Shu Q, Pasqua N, Outani O, et al. Mortality from gastrointestinal congenital anomalies at 264 hospitals in 74 low-income, middle-income, and high-income countries: a multicentre, international, prospective cohort study. Lancet (British Ed. 2021;398(10297):325–39.
- Adejuyigbe O, Abubakar AM, Sowande OA, et al. Experience with anorectal malformations in Ile-Ife, Nigeria. Pediatr Surg Int. 2004;20(11):855-8.
- Mohammed M, Amezene T, Tamirat M. Intestinal Obstruction in Early Neonatal Period: A 3-Year Review Of Admitted Cases from a Tertiary Hospital in Ethiopia. Ethiop J Health Sci. 2017;27(4):393–400.
- 26. Hasan MS, Islam N, Mitul AR. Neonatal Surgical Morbidity and Mortality at a Single Tertiary Center in a Low- and Middle-Income Country: A Retrospective Study of Clinical Outcomes. Front Surg [Internet]. 2022;9. Available from:

- https://www.frontiersin.org/article/10.3389/fsurg.2022.81752
- Levitt MA, Pena A. Anorectal malformations. Orphanet J Rare Dis. 2007;2(1):33.
- 28. Shakeel F, Newkirk M, Sellers A, Shores DR. Postoperative Feeding Guidelines Improve Outcomes in Surgical Infants. JPEN J Parenter Enteral Nutr. 2020;44(6):1047–56.
- Penman G, Tavener K, Hickey A. Neonatal feeding: care and outcomes following gastrointestinal surgery. Infant J. 2017;13(2):61-4.
- 30. Ballot DE, Bandini R, Nana T, Bosman N, Thomas T, Davies VA, et al. A review of multidrug-resistant Enterobacteriaceae in a neonatal unit in Johannesburg, South Africa. BMC Pediatr. 2019;19(1):320–9.
- Nkwanyana NM, Singh R, Mzimela BW. Clinical outcome of neonates with Carbapenem-resistant Enterobacteriaceae infections at the King Edward VIII Hospital's neonatal unit, Durban, South Africa. South African J Infect Dis. 2021;36(1):e1-6.
- 32. Thomas R, Ondongo-Ezhet C, Motsoaledi N, Sharland M, Clements M, Velaphi S. Incidence and All-Cause Mortality Rates in Neonates Infected With Carbapenem Resistant Organisms. Front Trop Dis [Internet]. 2022;3. Available from: https://www.frontiersin.org/article/10.3389/fitd.2022.832011 (accessed 20 June 2022)
- 33. Folgori TL, Bielicki TJ, Heath TP, Sharland TM. Antimicrobial-resistant Gram-negative infections in neonates: burden of disease and challenges in treatment. Curr Opin Infect Dis. 2017;30(3):281–8.
- 34. Shane AL, Sánchez PJ, Stoll BJ. Neonatal sepsis. Lancet (British Ed. 2017;390(10104):1770-80.
- 35. Mpody C, Shepherd EG, Thakkar RK, Dairo OO, Tobias JD, Nafiu OO. Synergistic effects of sepsis and prematurity on neonatal postoperative mortality. Br J Anaesth BJA. 2020;125(6):1056-63.
- Shetty S, Kennea N, Desai P, Giuliani S, Richards J. Length of stay and cost analysis of neonates undergoing surgery at a tertiary neonatal unit in England. Ann R Coll Surg Engl. 2016;98(1):56–60.
- Gasparella P, Singer G, Kienesberger B, Arneitz C, Fullop G, Castelleni C, et al. The Financial Burden of Surgery for Congenital Malformations—The Austrian Perspective. Int J Environ Res Public Health [Internet]. 2021;18(11166):11166-Available from: https://doi.org/10.3390/ijerph182111166 (accessed 28/06/2022)

- Lawal TA. Overview of Anorectal Malformations in Africa. Front Surg. 2019;6.
- Collaboration PAR. Paediatric surgical outcomes in sub-Saharan Africa: a multicentre, international, prospective cohort study. Wright NJ, Smith ER, Bisquera A, John-Chukwuemeka AL, Lawal TA, Seyi-Olajide J, et al., editors. BMJ Glob Heal [Internet]. 2021;6(9). Available from: https://gh.bmj.com/content/6/9/e004406
- Sholadoye TT, Mshelbwala PM, Ameh EA. Presentation and outcome of treatment of jejunoileal atresia in Nigeria. African J Paediatr Surg. 2018;15(2):84–7.
- 41. Du Preez H BE. Findings from the University of the Free State Yields New Data on Gastroenterology (The Profile and Outcome of Small Bowel Atresia At Universitas Academic Hospital). South African J Surg. 2023;61(1).
- Tarcă E, Cojocaru E, Trandafir LM, Luca AC, Tiutiucă RC, Butnariu LI, et al. Current Challenges in the Treatment of the Omphalocele—Experience of a Tertiary Center from Romania. J Clin Med. 2022;11(19):5711.
- 43. Rode H, Millar AJW. Our surgical heritage: the role of the Department of Paediatric Surgery in the development of paediatric surgery in Cape Town, in Africa, and around the world. South African Med J. 2012;102(6):409–11.
- Okoye MT, Ameh EA, Kushner AL, et al. A Pilot Survey of Pediatric Surgical Capacity in West Africa. World J Surg. 2015;39(3):669–76.
- 45. Burgos CM, Frenckner B. Addressing the Hidden mortality in CDH: A population-based study. J Pediatr Surg. 2016;52(4):522–5.
- Roorda D, Königs M, Eeftinck Schattenkerk L, van der Steeg L, van Heurn E, Oosterlaan J. Neurodevelopmental outcome of patients with congenital gastrointestinal malformations: a systematic review and meta-analysis. Arch Dis Child Fetal Neonatal Ed. 2021;106(6):635–42.
- 47. Stolwijk LJ, Lemmers PMA, Harmsen M, Groenendaal F, de Vries LS, van der Zee DC, et al. Neurodevelopmental Outcomes After Neonatal Surgery for Major Noncardiac Anomalies. Pediatrics [Internet]. 2016;137(2):e20151728-e20151728. Available from: https://pediatrics.aappublications.org/content/137/2/e20151728 (Accessed 07/10/2019)
- 48. Bevilacqua F, Ravà L, Valfrè L, Braguglia A, Zaccara A, Gentile S, et al. Factors affecting short-term neurodevelopmental outcome in children operated on for major congenital anomalies. J Pediatr Surg. 2015;50(7):1125–9.