Patterns and Short Term Outcome of Children with Major Congenital Anterior Abdominal Wall Defects Admitted At Mbarara Regional Referral Hospital, South Western Uganda, from September 2019 to June 2020.

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Abstract

Background: Omphalocele and gastroschisis are the most common major congenital abdominal wall defects (CAWDs) globally. Mortality among neonates major CAWDs is higher in low-income countries than in high-income countries. This study described the patterns and short-term clinical outcomes of infants with gastroschisis and omphalocele at a regional referral hospital in southwestern Uganda.

Methods: A case series was conducted at Mbarara Regional Referral Hospital ten months. Children born with CAWD during the study period 54 samples size were consecutively recruited into the study. Data were entered, cleaned, and analyzed using Stata version 15. Descriptive statistics were performed where continuous variables were summarized using median and interquartile ranges, while categorical variables were summarized using frequencies and proportions. Time to mortality was assessed using Kaplan–Meier survival analysis.

Results: A total of 54 newborns were recruited into the study, of which 40 (70.04%) had Gastroschisis and 14(25.93%) had omphalocele. The median age of the infants was 9 days with an interquartile range of 4 to 21days. Of the 54 newborns, 30(69.2%) were male within the age group of 1-10 days. Mortality was higher in babies with complex gastroschisis (22) than simple gastroschisis (10). Not administering antibiotics and low birth weight were the factors associated with mortality among infants diagnosed with gastroschisis and omphalocele in the neonatal period.

Conclusion: The most common observed defect was gastroschisis, Mortality was generally high among children presenting with gastroschisis. Complex gastroschisis has a poorer prognosis than simple gastroschisis over 30 days follow up period. Non-use of antibiotics before referral to a health facility and low birth weight were the factors associated with mortality among infants diagnosed with gastroschisis and omphalocele in the neonatal period.

Recommendation: The most common pattern observed was Gastroschisis. The majority of children presenting with Omphalocele were more likely to survive for 30 days. There is a need for timely referral for babies born with congenital anterior abdominal wall defects.

Keywords: *Congenital abdominal wall defect, Omphalocele, Gastroschisis, Mortality, Uganda*

Introduction

Major congenital abdominal wall defects (omphalocele and gastroschisis) are the most frequent foetal abdominal wall anomalies worldwide, with an estimated prevalence of 3 to 4 per 1,000 live births [1, 2]. Gastroschisis refers to full-thickness paraumbilical abdominal wall anomaly which is often associated with bowel evisceration, and in rare cases, evisceration of other abdominal organs. Omphalocele, on the other hand, is a midline defect of the abdominal wall that can vary in size, covered by a membranous sac[3]. Marked variation in the prevalence of Gastroschisis has been noted, with the reported rates being higher in single pregnancies than twin pregnancies [4].

Gastroschisis is associated with numerous risk factors, which include young maternal age, poor diet, being a primigravida, and cigarette smoking or use of certain drugs[5]. For the case of omphalocele, maternal age of 40 years or older, maternal obesity, and black race, have been implicated[5]. However, unlike gastroschisis, omphalocele is also linked to chromosomal abnormalities, including Beckwith-Wiedemann syndrome, Trisomy 21, 18, and 13[6, 7].

The prognosis and management of omphalocele and gastroschisis is largely determined by the complexity of abnormalities based on the existence of complications including necrosis, atresia, or gut perforation; presence of other congenital abnormalities or co-existing chromosomal abnormalities are other prognostic markers[8]. In many low-income countries, the reported mortality associated with these congenital abdominal wall defects (CAWDs) ranges from 30–100%, while in high-income countries, the mortality is less than 5%[9]. The mortality can go as high as 88% in omphalocele, when associated with congenital cardiac abnormalities; if there are no cardiac abnormalities, up to 70% of the cases survive [10]. Nevertheless, the high mortality rates reported in resource-limited settings compared to resource-rich countries provoke deep concern. In central Uganda, for instance, up to 100% mortality was reported among newborns with gastroschisis (Ford et al., 2016)However, the patterns and outcomes of the congenital anterior abdominal wall defects, among infants admitted in other regions in Uganda, including southwestern Uganda have not be characterized. In order to generate enough evidence to inform evidence-based management protocols for children with CAWDs in Uganda and other similar resource-limited settings, there is need to comprehensively assess clinical patterns and outcomes of the CAWDs in the various settings. This study assessed the patterns and clinical outcomes of children admitted at the surgical ward of Mbarara Regional Referral Hospital (MRRH) in southwestern Uganda.

Methods

Study Setting and Study Design

This was a descriptive case series conducted at Mbarara Regional Referral Hospital (MRRH) for 10 months from September 01, 2019 to June 30, 2020. The hospital has a 350-bed capacity. It is a government referral hospital for the southwestern region of Uganda. The study was conducted at the pediatric surgery unit of the Department of Surgery of MRRH. The Pediatric Surgical Ward has 15 beds and 3 incubators. There are currently four pediatric surgeons in the whole of Uganda, of whom one is based in MRRH. The hospital is located in Mbarara City, approximately 286 kilometers from Kampala, the capital city of Uganda.

Study Population

The study population included babies born with gastroschisis, and omphalocele and whose parents gave informed consent and excluded those who presented to the facility after 30 days post-delivery.

Recruitment of Participants and Sampling Procedures

The study team comprised two nurses working in the paediatric surgery ward and the principal investigator. On each working day, the study team conducted health education sessions at the paediatric surgical clinic ward for the admitted clients that day. The study team used this opportunity to sensitize mothers about the study and study procedures. The research assistants were stationed in the consultation rooms. After consultations, the potential study participants who fulfilled the study inclusion criteria were identified. Informed consent was obtained from the parents of the selected study participants prior to recruitment into the study. All participants who met the eligibility criteria during the study period were enrolled into the study, through consecutive sampling, and followed up for 30 days.

Data Collection Procedures, Study Variables and Definitions

Data were collected using interviewer-administered questionnaires, in a private study room to offer privacy. They socio-demographic data of the new-borns (age at admission, sex, birth weight, and place of birth), maternal characteristics (age, occupation, level of education, antenatal care attendance, marital status, and medication use during pregnancy). The diagnoses of gastroschisis and omphalocele were made by the attending paediatric surgeon. They defined complex gastroschisis in children with gastroschisis and intestinal complications including necrosis, atresia, or perforation; children with gastroschisis in absence of the complications were considered to have simple gastroschisis[11, 12]. The short-term outcome of interest was mortality in the cohort.

Data Management and Analysis

Data were entered and cleaned in Microsoft Excel, then analyzed using Stata version 15 (StataCorp, College Station. Texas, US). Descriptive statistics were performed where continuous variables were summarized as medians (interquartile ranges); while categorical variables were summarised by frequencies and percentages of occurrence. Differences between categorical socio-demographic and clinical characteristics of children with gastroschisis and those with omphalocele were assessed using chi-square or two-sided Fischer's exact tests. Wilcoxon rank-sum test was used for comparing non-normally distributed continuous variables. Time to mortality (our outcome of interest) was assessed using Kaplan–Meier survival analysis. The follow-up time was 30 days (from admission). The probabilities of survival between the two groups (omphalocele and gastroschisis) were compared using the Log-rank test. A p value<0.05 was considered statistically significant.

Results

Baseline Characteristics of Children with Omphalocele and Gastroschisis

A total of 54 children with omphalocele and gastroschisis were recruited during the 10 months study period. Of the 54 children, 38 (70.4%) were male newborns; 40 (74.1%) had gastroschisis. Of the 40 children with gastroschisis, 14 (35%) had complex gastroschisis. The median age was 9 days (IQR: 4-21) days. Most (56.4%) of the children weighed 1.6 - 2.5 kgs as shown in table 1.

Table 1: Baseline characteristics of children by omphalocele and gastroschisis abdominal wall defects

Characteristic	Overall (N=54) n/N (%)	Gastroschisis (n=40) n/N (%)	Omphalocele (n=14) n/N (%)	P value
Age in days, median (IQR)	9 (4 -21)	10 (4-21)	6 (4-9)	0.657
Age in days				0.105
1-10	31(57.4)	21(52.5)	10 (71.4)	
11-20	9 (16.7)	9 (22.5)	0 (0.0)	
21-30	14(25.9)	10 (25.0)	4 (28.6)	
Birth weight in kgs				0.037
≤1.5 kgs	11 (20.4)	7 (17.5)	4 (28.6)	
1.6 - 2.5 kgs	33(61.1)	27 (67.5)	6 (42.9)	
>2.5 kgs	10 (18.5)	6 (15.0)	4 (28.6)	
Sex of baby				0.602
Male	38 (70.4)	29 (72.5)	9 (64.3)	
Female	16 (29.6)	11 (27.5)	5 (35.7)	
Place of birth				0.570
Home	18 (33.3)	12 (30.0)	6 (42.9)	
Health Centre	16 (29.6)	12 (30.0)	4 (28.6)	
Hospital	20 (37.0)	16 (40.0)	4 (28.6)	
Received antibiotics				0.021
Yes	37 (68.5)	23 (57.5)	14(100)	
No	17(31.5)	17 (42.5)	(0.0)	
Defect size				0.330
0-3 cm	9 (16.7)	8 (20.0)	1 (7.1)	
3-6 cm	35 (64.8)	22(55.0)	13 (92.9)	
7-15 cm	10 (18.5)	10 (25.0)	0 (0.00)	

IQR: Interquartile range

Mortality among Infants with Gastroschisis and Omphalocele

Of the 54 infants, 30 died before the 30th day of admission, for an overall mortality rate of 55.6%. Of the 40 children with gastroschisis, 27 (67.5%) died, compared to 3/14 (21.4%) of infants diagnosed with omphalocele. Survival time was significantly higher among children with omphalocele compared to those with gastroschisis (*P* value=0.038) as indicated in figure 1. The probability of surviving for 30 days' (720 hours) follow-up among the children who presented with gastroschisis was 0.25 while the probability of surviving for 30 days (720 hours) of follow-up among those with omphalocele was 0.78.
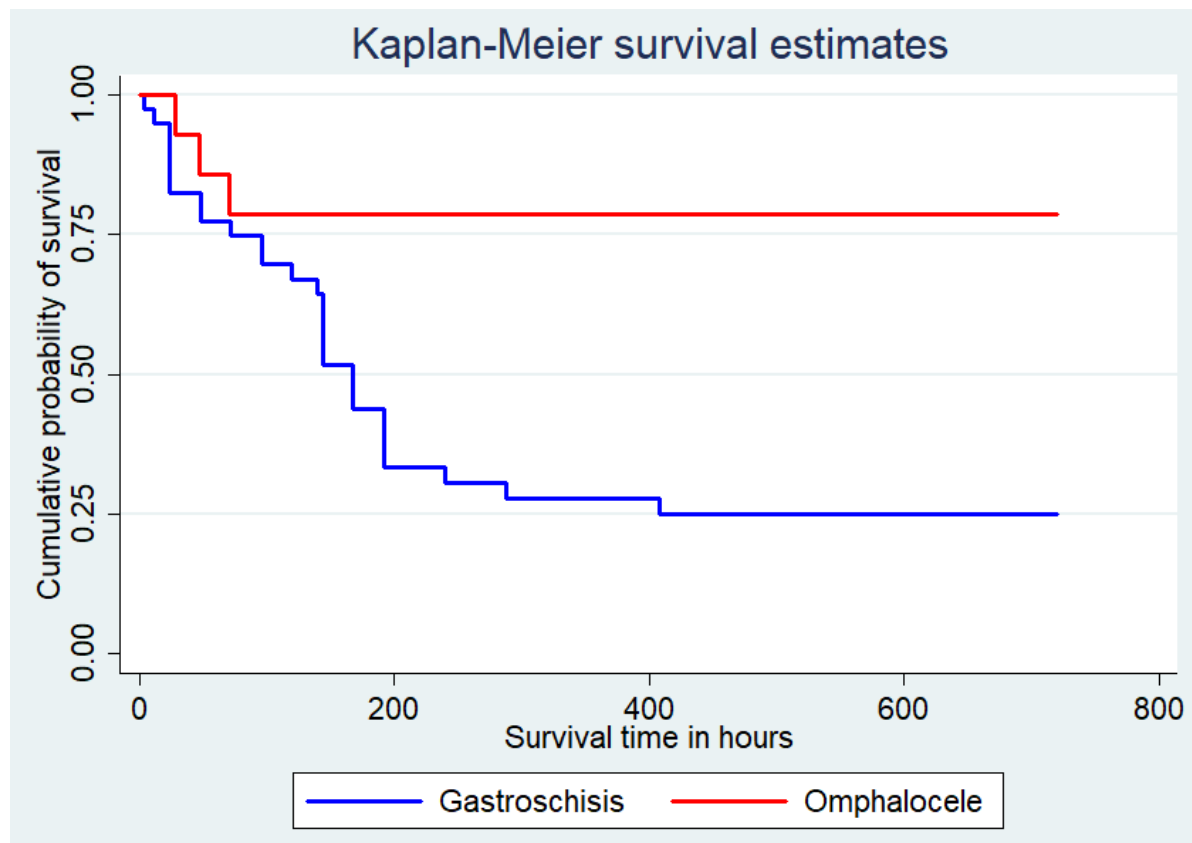
Figure 1. Survival rates of children with gastroschisis and omphalocele admitted at Mbarara Regional Referral Hospital, southwestern Uganda, from September 2019 to June 2020

Children presented with gastroschisis died within the first 100 hours of life as shown in figure 2.
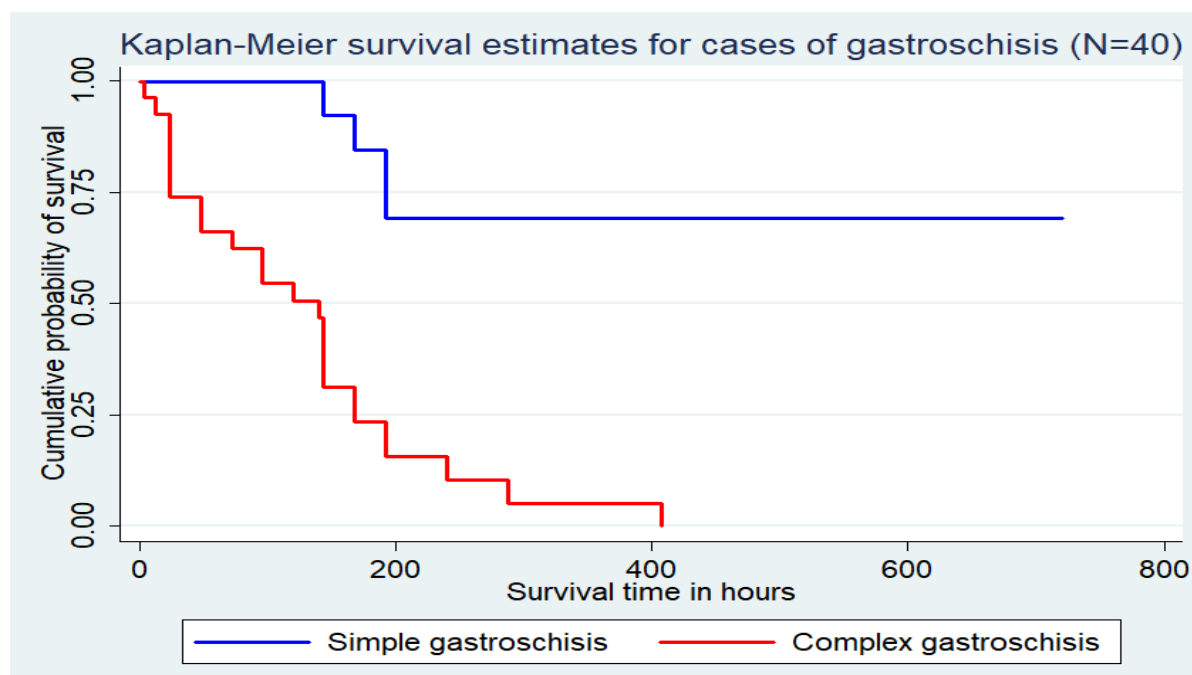

Figure 2: Kaplan-Meier survival estimates for gastroschisis.

The probability of surviving for 30 days (720 hours) follow-up among those children who presented with simple gastroschisis was 0.65 while the probability of surviving for 30 days (720 hours) of follow-up among those with complex gastroschisis was 0.01 (*P* value=0.001); all participants with complex gastroschisis did not make it to 720 hours (30 days). Results from this study show that of the children with Gastroschisis abdominal wall defect who live for at least 18 days make to 30 days and that those suffering from omphalocele abdominal wall defect after living for 8 days make 30 days. Furthermore, results revealed that the 30-day outcome of children with Gastroschisis of the referred cases at MRRH: 67.5% died while 32.5% had successful repairs. In the same way, the outcome of children with omphalocele abdominal wall defect referred cases at MRRH: 14.2% died while 85.8% successfully made to 30 days.

With regards to Gastroschisis prognostic score, GPS enables risk stratification for Gastroschisis and helps discriminate low from high morbidity groups children with higher GPS had a poor prognostic outcome than children with low GPS this is in agreement with a study by puligandla on outcome predictors for babies born with Gastroschisis(Puligandla et al., 2017)

Conclusion

The most common pattern observed was Gastroschisis. The majority of children presenting with Omphalocele were more likely to survive for 30 days. There is a need for timely referral for babies born with congenital anterior abdominal wall defects. Simple Gastroschisis has a better prognosis than complex Gastroschisis (Anyanwu et al., 2020).

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