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Gastroschisis: A multi-centre comparison of management and outcome

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ABSTRACT

Background: Anecdotal evidence and a handful of literature reports suggest that the outcome for infants born with gastroschisis in many African countries is poor when compared to Western nations. We wished to evaluate current management strategies and outcomes in African and Western units that treat infants with gastroschisis. **Patients and Methods:** We conducted a retrospective review of case-notes for infants with gastroschisis who presented to a hospital between 1 January 2004 and 31 December 2007. There were five participating centres, divided for analysis into an African cohort (three centres) and a Western cohort (two centres). **Results:** Fewer infants presented to a hospital with gastroschisis in the African cohort when compared to the Western cohort, particularly when the size of catchment area of each hospital was taken into account. The physiological state of the infant on presentation and management strategy varied widely between centres. Primary closure, preformed silo and surgical silo with delayed closure were all utilised in the African cohort. Use of the preformed silo and delayed abdominal wall closure was the strategy of choice in the Western cohort. The 30-day mortality was 23% and 1% respectively. This primary outcome measure varied considerably in the African cohort but was the same in the two Western units. **Conclusions:** Gastroschisis in the African cohort was characterised by fewer infants presenting to a hospital and a more variable outcome when compared to the Western cohort. A detailed epidemiological study to determine the incidence of gastroschisis in African countries may provide valuable information. In addition, interventions such as prompt resuscitation, safe neonatal transfer, the use of the preformed silo and parenteral nutrition could improve outcomes in infants with gastroschisis.

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INTRODUCTION

Gastroschisis (GS) is a congenital condition, characterised by a full-thickness defect in the anterior abdominal wall, with intestinal herniation. It is strongly associated with young maternal age[1] and is usually an isolated defect.[2]

The survival of infants with GS in the UK is reported to be over 90%[3] and long-term cohort studies of infants who survived beyond one year, suggest that 96% had normal growth and normal health although 35% required further surgery.[4] By contrast, literature from Africa has demonstrated poorer outcomes and wide variation between centres.[5-8] A number of strategies that may influence outcomes are used in the management of this condition and these may be dictated by institutional history, staff capacity, preference of the surgeon, and availability of facilities and materials. We wished to evaluate management strategies and clinical outcomes in a comparative series of infants with GS managed in African and Western units.

MATERIALS AND METHODS

Five paediatric surgery units participated. A retrospective case-note analysis on all infants with GS presenting to the institutions between 1 January 2004 and the 31 December 2007 was performed. Each centre maintains an up-to-date, comprehensive electronic or paper database of paediatric surgery procedures. The databases allowed identification of infants who presented with GS. The centres were divided into two cohorts for analysis: Western cohort (WC) and African cohort (AC).

The materials used in this study were delivered by Jo Manson for which she won the prize for the best oral presentation by a trainee at the Pan African Paediatric Surgical Association (PAPSA) Congress in Accra, Ghana, 2008.

The medical records were reviewed to extract relevant data. A standardised spreadsheet (Microsoft Excel®), with an accompanying explanatory document ensured uniformity of data collection. The information recorded included the date of admission, gestational age, birth-weight, presence of intestinal peel or associated anomalies, the condition of the baby on arrival and the timing and type(s) of surgical intervention. Preformed silo (Bentec® and Medicina®) application was carried out in standardised fashion in the two units where it was routinely used. Outcomes and complications were recorded. Except where explicitly stated, data are reported as median with ranges.

RESULTS

All the participating centres were urban teaching hospitals with three to five Paediatric Surgery Consultants. The catchment areas were larger for the African centres. The capacity of each institution varied in terms of the number of paediatric and neonatal intensive care (NICU) beds. During the study period, prenatal sonography, intravenous nutrition and intensive care facilities were only available in London, Irvine, and Cape Town. The number of infants presenting to each hospital was similar for the Western units; London 59, Irvine 58, but the African sites showed a wide variation; Accra 2, Zaria 5, Cape Town 19 [Table 1].

All infants in the WC (n=110) were diagnosed with GS during routine antenatal ultrasound. Once a foetal diagnosis of GS was made, antenatal care was transferred to a specialist foetal medicine unit at the tertiary care institution. In California, all infants were delivered either at the University of California Irvine (UCI) Medical Center or at Miller Children's Hospital, a tertiary teaching hospital affiliated with UCI. In London, elective induction at 38 weeks was arranged but spontaneous labour prior to this date led to some early *in*- and *ex-utero* transfers to King's College Hospital for management. All WC infants were admitted to the Tertiary Centre within 24 h of delivery.

In the AC (n=26), 7.7% had an antenatal diagnosis. All infants were delivered after spontaneous labour. The majority (n=16) presented within 24 h however, nine presented on Day 2 and one presented on Day 3.

In the WC, the birth weight was 2480 g (1170–3904) and the gestational age 37 weeks (30–9). Eighty per cent were born in good condition, 9.6% were "septic", 3.6% required early ventilation, and 1.8% had ischemic bowel at delivery. In the AC, the birth weight was 2200 g (1750–3300) and the gestational age 36 weeks (32–40). The condition of the infants varied depending on the geographical location of the centre. In Cape Town (n=19), 79% of the infants were born in good condition, 5% had "pneumonia" and 16% required ventilation. In Zaria, 60% were hypothermic, 20% were "septic" and 20% were both.

In the WC (n=110) 25 infants (23%) had a primary closure on Day 1. In the London cohort there were no primary closures after 2004 due to a departmental move towards preformed silos (PFS). Eighty-four infants had a PFS fitted on Day 1 with delayed closure on Day 6 (3-16) of life; 8.3% of the PFS were placed under general anaesthetic, as minor surgery was required to divide skin or mesenteric attachments, free the bowel or widen the defect.

One infant had the defect closed using steri-strips in the neonatal unit on Day 7 and the remainder were closed in theatre. The majority underwent simple closure (79%), however, 11% required more complicated procedures, which included patch repair (one), bowel resection (three) and a Ladds Procedure (one). One had a surgical silo (SS) with closure on Day 4. The infants reported as "Closed" GS underwent a primary closure in six out of the eight cases. Of those other two, one had an SS and one had a PFS placed in theatre after widening of the defect [Figure 1].

In the AC (n=26), two infants had a PFS and delayed closure on Days 2 and 8. Twelve had a primary closure on Day 1, 10 had an SS placed and delayed closure on Day 4.5 (3-13) and two were considered unfit for surgery, so had no intervention. In the PFS group one had a Simple closure, one had a resection and closure. In the primary closure group, one required a Ladds Procedure for malrotation. The SS group was more complicated: Simple closure (one), laparotomy and closure (one), laparotomy and resection (one), laparotomy and resection but not closed (one), laparotomy and reversal of ileostomy D4 (one) (ileostomy formed on Day 1; infant had malrotation and underwent caecal resection with ileocolic anastomosis

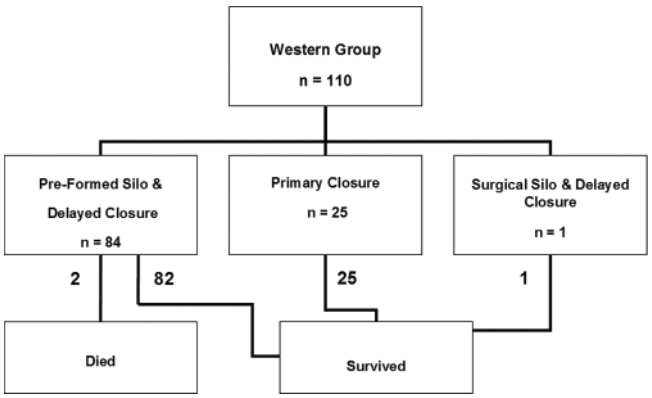


Figure 1: Management and outcome of the Western cohort

Table 1: Summary of demographics, centre capacities and physiological condition at presentation

	London, UK	Irvine, USA	Accra, Ghana	Zaria, Nigeria	Cape Town, SA
Centre capacity					
Estimated catchment area (millions)	3	2	6	35	4.6
Number of consultant staff	4	3	3	4	5
Paediatric surgical beds	20	25	47	16	62
NICU beds	18	95	0	0	12
Gastroschisis infants in study					
Total infants who presented (n)	59	58	2	5	19
Male (%)	46	64	-	40	47
Antenatal diagnosis (%)	100	100	0	0	7.7
Presented on day of delivery (%)	92	100	0	80	63
Present next day (%)	8	-	100	20	32
Present > 24 hrs (%)	-	-	-	-	5
Birth weig ht (g)	2480 (1170-3940)			2200 (1750-3300)	
Gestation (weeks)	37 (30-39)			36 (32-40)	
Physiological condition at presentation					
Good condition (%)	81	45	-	-	79
Sepsis and hypothermia (%)	0	0	-	20	-
Sepsis alone (%)	8	55	-	20	-
Hypothermia alone (%)	0	0	-	60	-
Pneumonia (%)	0	0	-	-	5
Required intubation (%)	11	0	-	-	16
Associated anomalies					
Turners syndrome (%)	0.9				
Intestinal atresia (%)	8.1			3.8	
Cryptorchidism (%)	5.5				
Closed gastroschisis (%)	7.3				
Hydronephrosis (%)				3.8	
Malrotation (%)				15.4	
Colonic duplication cyst (%)				3.8	
Outcomes					
30 day mortality (%)	1			23	
Total length of stay (days)	33 (15-217)			24 (8-121)	
Transferred to home (n)	16	56	2	0	10
Transferred to local hospital (n)	35	2	0	0	6
IVN dependant at discharge (n)	4	0	0	0	3

Data stated as number (*n*) or percentage within the cohort (%) or median (with ranges)

and defunctioning ileostomy) [Figure 2].

In the WC, the 30-day mortality was 1% as one infant died from multiple congenital abnormalities on Day 4. One further death occurred secondary to sepsis on Day 76. In the AC, the 30-day mortality was 23% (n=6). However, there were considerable differences in outcomes between institutions; the 30-day mortality rate was 100% at Zaria, 50% in Accra and 5.2% in Cape Town. Four infants died within the first 48 h of life due to: Sepsis and multiple organ failure (one), hypothermia (one) and respiratory insufficiency (two). One died on Day 9 from cardiorespiratory arrest following a pneumothorax, another on Day 21 although there is insufficient information to determine the cause. One SS infant was lost to follow-up after his parents took him from the hospital on Day 5. One further death occurred on Day 43, due to sepsis and multiple organ failure [Figure 3].

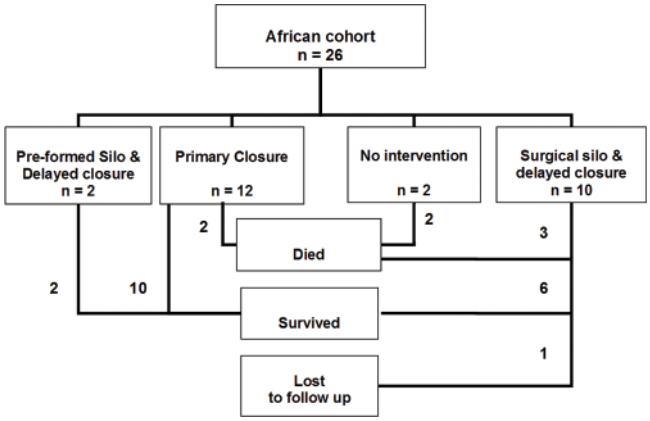


Figure 2: Management and outcome of the African cohort

In the WC (n=108), five infants (6%) experienced problems with the PFS (n=84). This included the need to widen the defect in two patients on Days 4 and 6, at

which time, another silo was placed. Ischemic pressure of the anterior duodenum from the silo ring occurred in two patients with complicated GS (one with multiple atresia, one with atresia and *in utero* volvulus). After closure, morbidity included necrotising enterocollitis 0.9%, "sepsis" 9.3%, wound infection 3.7% and hyperbilirubinaemia due to intravenous nutrition 3.7%. Eight infants underwent a further laparotomy (7.4%). One of the infants was referred for and was accepted for, a bowel transplant programme.

In the AC (n=18) morbidity included: PFS falling off 100% (n=2), GI bleed 5.6%, overt compartment syndrome 11.1%, sepsis 28%, enterocutaneous fistulae formation 5.6% (n=1) and bowel perforation 5.6% (n=1). These last two infants underwent multiple operations including repeated laparotomy and stoma formation. Of the infants given intravenous nutrition (n=19), 25% required more than one surgically placed central line [Figure 4].

Intravenous nutrition (IVN) was used for 25 days (7–05) in the WC and 18 days (6–111) in the AC (Cape Town only). The total length of stay was 33 days (15-217) in the WC and 24 days (8-121) in the AC. In the WC, four infants were still IVN-dependent at discharge. In London, 35 infants were transferred to a local hospital to facilitate local medical support, one destination was unknown and 16 were discharged home. In UCI, 56 went home at discharge and two were transferred to a local hospital. In the AC, three patients were still IVN-dependent at discharge from tertiary hospital. Six were transferred to their local hospital and 12 were discharged home.

DISCUSSION

The incidence of GS is increasing in many countries.[9,10] There is limited representation of African data in the literature and currently the incidence of GS in many countries in Africa is not quantified. Arnold looked at the occurrence of GS as a proportion of the paediatric surgical admissions to two hospitals in Pretoria in South Africa over a 21-year period.[11] Consistent with the worldwide experience, a 35-fold increase from 1981–1987 compared to 1995–2001 was demonstrated. A striking feature of this current study is the relatively small numbers of infants with GS in the AC, considering the size of the hospital catchment areas. For example, The Korle Bu Hospital is one of two paediatric surgery units in Ghana and has a catchment population in the region of 6 million yet only two infants were treated during the study period. Assuming an incidence of three per 10,000 live births and using a UNESCO-validated fertility rate of 33.4 per 1,000 of the Ghanian population, 53 infants with GS should have been born during the study period. It is unclear whether the low figures in this report represent the true incidence of this birth defect. Anecdotal reports and personal communication with a range of African health professionals suggest that some infants born with GS do not reach a hospital. Further epidemiological research is required.

The physiological state of the infant at presentation is predictive of outcome. In 2006, Harrison estimated that for infants with GS in Zimbabwe, every hour of delay between their delivery and hospital treatment results in an 8% increased risk of mortality.[6] In Cape Town, the majority of infants presented in good condition and their mortality was lower. By comparison, the typical infant presenting in Zaria was in extremis, reflecting the long distances travelled with inadequate resuscitation and insufficient prevention of fluid and heat loss. Not surprisingly, the mortality was high. Molik[2] has previously called for GS risk categorisation to allow meaningful comparisons of outcomes within series and between centres. In this report and in a number of others, high morbidity and mortality related to poor physiological state, particularly systemic and intestinal hypoperfusion is recognised. Furthermore, the importance of avoiding abdominal compartment syndrome as an additional insult in GS is increasingly well recognised.[12]

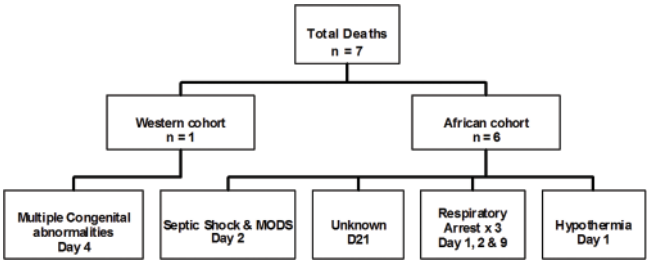


Figure 3: Thirty-day mortality

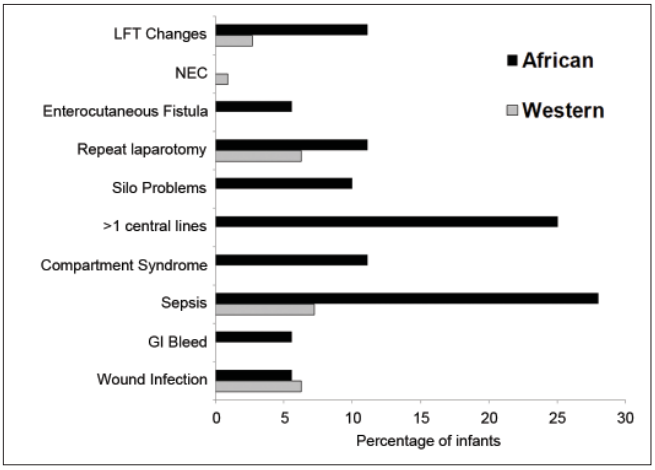


Figure 4: Morbidity of the survivors in both cohorts

Improving the physiological condition of the infants at presentation is the key to improving outcome and this could be supported by education at each stage. Teaching traditional birth assistants (TBAs) and other primary care providers to "Recognise, Resuscitate and Refer" may play an important role. At the receiving hospital, further resuscitation and stabilisation is imperative. Surgical options can then be considered once the patient is stable.

Predominantly, the WC infants had a PFS placed on Day 1 and the indication to proceed to theatre was often failure to place a PFS. In the AC, 84% of the infants underwent an operative procedure under general anaesthetic on the first day of arrival compared with 30% in the WC. Infants predominantly underwent primary closure or SS as PFS were only available in Cape Town and only used in two infants. It may be that more widespread use of the PFS would enable the infants to stabilise physiologically before undergoing a general anaesthetic. For surgeons with limited or no previous experience, use of an *ex-vivo* training model could reduce the time taken to achieve expert competence in the application of the PFS.[13]

The effect of concurrent HIV infection on morbidity or mortality of GS infants is not known. We did not record HIV status as part of our study but it may account for the higher rate of sepsis and the development of conditions such as enterocutaneous fistulae, which were unique to the AC.

Postoperative feeding was intravenous nutrition (IVN) in 94% of the infants at the institutions which had IVN available. At Accra and Zaria oral feeding was the only available route. In 2000, Ameh reported "no adverse affects from the early introduction of oral feeds"[5] but those patients may have been a self-selected group who survived despite difficult circumstances.

This report highlights the challenges faced by African paediatric surgeons when treating neonatal surgical pathology with limited facilities and resources. A difference in the management and outcomes of GS infants between African and Western centres has been demonstrated. In the WC, a multidisciplinary team provides care with a consistent strategy and achieves consistently good outcomes. In the AC, there are far fewer infants than expected and the treatment strategy varies according to the physiological condition of the infant and the facilities available. Reducing the mortality of GS in African infants is likely to require additional resources for neonatal care and nutrition. International collaboration could help to expedite this.[14,15]

Two infants in the AC presented in extremis and died within 24 h. Another was lost to follow-up; after a surgical silo had been placed the parents elected to take the child from hospital against medical advice. A higher rate of complications was reported in the AC and these were of a diverse nature. PFS was only used on two infants but both had problems related to silo retention. Enterocutaneous fistulae were peculiar to the African group and where central lines were used, the sepsis rate was high. Concerted local and international efforts could help improve the outcomes of infants in Africa with GS so that morbidity and mortality more closely approximate international benchmarks.

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