Omphalocoele and gastroschisis: management in a developing country

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Abstract

Background: The management of omphalocoele and gastroschisis presents a lot of challenges. While most of these challenges have largely been overcome in developed countries, the same cannot be said of the developing countries.
Methods: The medical records of neonates with omphalocoele and gastroschisis seen at Jos University Teaching Hospital between January 1991 and December 2001 were retrospectively reviewed.
Results: There were 42 neonates with omphalocoele and 12 with gastroschisis. The median age at presentation of neonates with omphalocoele with intact sac was 72 hours (range 1-5 days) while the median age for those with ruptured sac was 14 hours (range: 3-36 hrs). The median age at presentation of neonates with gastroschisis was 10 hours (range: 2-40 hrs.) Six neonates with ruptured omphalocoele and 4 with gastroschisis had gangrenous bowel at presentation. Eighteen neonates with omphalocoele had associated congenital anomalies, mostly involving the gastrointestinal and genitourinary systems. No neonate with gastroschisis had associated congenital anomaly. Sixteen neonates with omphalocoele were managed non-operatively, while 26 had operative treatment at initial presentation. Of these, 14 had primary fascial closure, 8 had skin closure, and 4 had improvised silo. All neonates with gastroschisis had primary fascial closure at presentation. A total of 22 neonates died: eighteen omphalocoele (11 of them managed surgically) and 4 gastroschisis.
Conclusion: The management of neonates with omphalocoele and gastroschisis continues to pose challenges in our environment, with high mortality rate. Health education, provision of neonatal intensive care facilities, availability of total parenteral nutrition in ours, and similar environment may improve the survival rate of this group of patients.

Key words: Omphalocoele, gastroschisis, developing countries

Introduction

The survival of neonates with omphalocoele and gastroschisis has improved greatly in developed countries over the year. [1, 2] In developing countries, the situation is different. [3,4] The impressive survival in the developed countries is largely due to the availability of total parenteral nutrition (TPN) and neonatal intensive care units. [1, 2] These facilities are not readily available in many developing countries. This is a report of a recent experience with the management of omphalocoele and gastroschisis in a developing country.

Patients and methods

The medical records of all patients with ompholocoele and gastroschisis managed at the Jos University Teaching Hospital between January 1991 and December 2001 have been retrospectively reviewed. Data were obtained from operation register, operation notes, discharged summary sheets and case records. Omphalocoele was defined as herniation of some of the intra-abdominal contents through the open umbilical ring into the base of the umbilical cord, while gastroschisis is full thickness abdominal wall defect lateral to a normally inserted umbilical cord. Omphalocoele major was identified as a defect with a base diameter >5cm while minor, a base diameter <5cm. A total of 57 neonates were identified with omphalocoele and gastroschisis, but 54 had adequate records and formed the basis of this report.

Results

There were 54 neonates, 42 (77.8%) of who had omphalocoele while 12 (22.2%) had gastroschisis.

Omphalocoele

Twenty-seven (64.3%) were boys while 15 (35.7%) were girls (M: F 1.8:1). Thirty were delivered outside the Jos University Teaching Hospital, by traditional birth attendants (TBAs). In 23 neonates, pregnancy was supervised in antenatal clinics. None was diagnosed antenatally. All deliveries were normal. The median age at presentation was 14 hours. (range 3-36 hrs) for those with ruptured sac, and 72 hours (range 1-5 days) in those with intact sac. Thirty-four (81%) of the omphalocoeles were major while 8 (19%) were minor. The contents of the omphalocoele in those that had surgery included intestines in all of them, additional liver and gall bladder in 5, and stomach in 2 patients.

At presentation, the sac was intact in 29 (69.0%) and ruptured in 13 (31.0%). All the ruptures occurred after delivery. In 6 neonates, the eviscerated bowels were gangrenous at presentation. A total of 24 associated malformations were detected in 18 patients, all with omphalocoele major. These anomalies are shown in table 1.

The treatment is summarized in table 2. A total of 16 omphalocoeles with intact sac were treated non-operatively. This involved admission to hospital, application of topical escharotics, like 1% silver sulphadiazine or povidone iodine, until the skin cover of the sac was achieved, converting it into a ventral hernia. The reasons for the choice of the non-operative treatment included infected sac, very large defect and the surgeon's preference. The median duration of hospital stay for patients managed non-operatively was 27 days (range 21-56 days). They were then discharged for a later operation after 1 year. So far there had been only 2 patients that had had repair of ventral hernia, as the others were lost to follow up.

Twenty- six patients had surgical treatment at presentation. These included

13 were patients with ruptured sac and 13 others with intact sac. Neonates with ruptured sac were initially resuscitated by keeping them warm, dehydration and electrolyte deficits corrected, and nasogastric tube passed to decompress the stomach. We routinely gave all of them parenteral vitamin K, and covered the exposed bowel loops with warm gauze soaked in normal saline. In the intact sac, defects were closed in patients with omphalocoele minor (8 patients) and 5 others with omphalocoele major, but with impending rupture (2 actually ruptured a few hours to surgery). Closure was performed under general anaesthesia with full muscle relaxation. In 13 patients fascial closure was achieved after a manual stretching of the anterior abdominal wall. Six of these 13 patients also had bowel resection because of gangrenous intestines. In 8 neonates, only skin-flap cover was possible. The resultant ventral hernia has not been repaired in any of them, as the patients are lost to follow up. Six patients had improvised silo in form of sterile latex gloves (3) and collapsible infusion bags in (3). The outcome was not as good as in those managed non-operatively. Table III lists the postoperative complications in those treated operatively. Prolonged ileus, respiratory problems and sepsis were the main forms of morbidity. Eighteen (43%) neonates died: eleven in those managed operatively and 7 in the non-operative group. The causes of death are not known for sure since the parents declined to postmortem examination.

Gastroschisis

There were 12 cases of gastroschisis. Eight (67%) were boys while 4 (33%) were girls (M: F 2:1). The median age at presentation was 10 hrs (range 2-40 hrs) and median weight at presentation was 2.5 kg (range 2-3.6 kg). Ten were delivered outside the Jos University Teaching Hospital, by Traditional Birth Attendants (TBAs). Four (4) mothers had antenatal care, but none was diagnosed antenatally. All deliveries were normal. The defect was right- sided in all with a median diameter of 3cm (range 2-4cm). Four babies had gangrenous bowel at presentation. There were no associated congenital anomalies noted in any neonate. Table 2 summarizes the treatment, and table 3 summarizes the postoperative morbidity. Four neonates died postoperatively, all of who had gangrenous bowel at presentation.

Table I. Associated malformations in 18 neonates with omphalocoele major

Malformation	No.
Beckwith – Wiedeman syndrome	4
Cloacal exstrophy	4
Bowel malrotation	11
Downs Syndrome	1
Macroglossia	4

Table 2: Treatment of ompholocoele and gastrochisis in 54 neonates

Treatment	Omphalocoele	Gastroschisis	Total
Non-operative	16	-	16
Surgical			
Primary fascial closure	13	9	22
Skin flap closure	7	2	9
Improvised silo closure	6	1	7
Total	42	12	54

Table 3: Post- operative complications in children undergoing omphalocoele and gastroschisis repair

Complication	Omphalocoele (%) n=26	Gastroschisis (%) n = 12
Ileus >2wks	12 (46.2)	7 (58.3)
Atelectasis/respiratory embarrassment	4 (15.4)	4 (33.3)
Pneumonia	3 (11.5)	-
Wound infection	2 (7.7)	1 (8.3)
Sepsis	4 (15.4)	3 (25.0)

Discussion

The results of this review compare favorably with those in most reports. [5 - 10] Most of our patients were delivered at home, which could partly be responsible for the delayed presentation, however, those with ruptured omphalocoele and gastroschisis tend to present earlier, compared with those with intact sac. As a direct consequence of the late presentation, the babies were hypothermic, with severe electrolyte and fluid deficits after having arrived hospital over long distances, with the bowel exposed and often gangrenous. Some patients were septic at presentation, adding to the metabolic stress that they already had. These factors also noted in other reports, [4, 11 - 14] often lead to high morbidity and mortality. Survival from omphalocoele and gastroschisis has greatly improved in most developed countries, largely as a result of prenatal diagnosis, advances in neonatal intensive care, total parenteral nutrition and proper transportation of babies to tertiary health facilities. [1, 2, 5] These services and facilities, however, are not readily available in developing countries like ours.

The operative management of emphalocoele major and gastroschisis is controversial. Though primary fascial or skin closure can be achieved in must patients, [1, 2, 8] good results have also been obtained with silo method. [15] Ventilatory support is often needed in patients with large defects. In our patients, we managed large defects non-operatively because we do not have functional neonatal intensive care units with neonatal or infant ventilators. Because of lack of proper silastic material, we improvised silos with gloves or infusion bags. Though this has been reported elsewhere, [4] we lost all the patients that had such management. We have since abundant this method.

We employed conservative management in most of our patients with major omphalocoele with intact sac. Previous authors [3] in Nigeria had reported an impressive survival rate from omphalocoele major with the use of the conservative management. This consisted of generous panting of the sac with 1% silver sulphadiazine (flamazine) daily, or the painting of the sac with povidone iodine. We have not seen any patients with hyperthyroidism following the use of povidone iodine, though our patients are few and follow up is a major problem in our environment. The prolonged hospital stay reported by previous authors 3 following the conservative approach, was confirmed in the present series. The duration of hospital in this study ranged from twenty one to fifty six days, with a median of twenty- seven days. This definitely creates social and financial stress on our patients, majority of whom are low- income earners. The overall mortality of 43% is higher than that

reported from the developed countries. [1, 2, 8] Though the presence of major congenital anomalies tends to carry a poor prognosis, [16] other preventable factors enumerated above may be the major determinants of this high mortality in this present report. It is hoped that the provision of functional neonatal intensive care units, availability by parenteral nutrition, and early presentation to the hospital would improve the present mortality.

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