Original Article

Ruptured and Non-Ruptured Omphaloceles: Characteristics, Management and Mortality Risk Factors in a Nigerien Cohort

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Abstract

Background: Omphalocele is a congenital anterior abdominal wall defect classified by size and sac integrity. In resource-limited settings, ruptured forms, low birth weight and delayed referral are associated with high mortality, yet ruptured and non-ruptured forms are seldom compared directly. **Aim:** To evaluate the distinctive diagnostic, therapeutic, and prognostic aspects of ruptured and non-ruptured omphaloceles. **Method:** This prospective, descriptive study included all patients with omphalocele admitted to the Paediatric Surgery Department of the National Hospital Amirou Boubacar Diallo Niamey, Niger between 1st August, 2023 and 28th February, 2025 (19 months). Sociodemographic, diagnostic, therapeutic, and outcome variables were analysed. Patients with ruptured and non-ruptured omphaloceles were compared using the Mann–Whitney and chi-square tests and Student's t-tests ($p<0.05$ was considered significant). **Results:** Eighty patients were included (5.6% of all hospitalizations), and mean admission age was 1.76 days (median, 18 h). Regarding prenatal care, only 6.25% (n=5) of the patients attended ≥6 consultations, and 14.64% (n=6) had antenatal diagnoses. Patients with ruptured omphaloceles (41.25%, n=33) showed significantly lower birth weight ($p=6.54\times10^{-19}$), higher primigravid prevalence ($p=0.04$), and younger maternal age ($p=6.54\times10^{-19}$) than those with non-ruptured omphaloceles. Pre-referral management gaps included absent nasogastric tubes (97.5%, n=78), delayed intravenous access (76.25%, n=61), and inadequate sac protection (36.25%, n=29). Treatment included the Grob (60%, n=48) and Gross (18.75%, n=15) techniques. Complications (18.75%, n=15) were predominantly due to sepsis (n=6), and the mortality rate was 51.25% (n=41), which was due to sepsis (n=12) and respiratory failure (n=8). Ruptured omphalocele ($p<0.00000001$), low birth weight ($p=0.0039$), admission delay ($p=6.53\times10^{-19}$), and primigravidity status ($p=0.0237$) correlated with mortality. **Conclusion:** This study highlights the critical impact of ruptured omphalocele, low birth weight, and delayed admission on omphalocele mortality in Niger. The increasing incidence and clear causes of ruptured omphalocele remain to be explored.

Keywords: Newborn, Ruptured Omphalocele, Anterior abdominal wall defect, Niger

Introduction

Omphalocele is a congenital anterior abdominal wall defect, which is classified according to size and sac integrity.[1-4] Lack of prenatal diagnosis, late neonatal presentation, and limited access to adequate care can lead to major complications.[1,2,5] Conservative management with delayed surgical closure is often preferred for intact sacs, especially in resource-limited settings.[3,6] Studies in Sub-Saharan Africa report high mortality rates, particularly for ruptured omphaloceles and those with large defects or associated anomalies.[2-4,7] Between 2020 and 2023, we witnessed an increasing incidence of omphalocele in our department, which prompted this study. The objectives of this study were to describe the diagnostic and management characteristics of omphaloceles in a resource-limited setting, to compare ruptured and non-ruptured omphaloceles, and to identify factors associated with mortality among neonates with omphalocele.

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Methodology

This was a single-centre, prospective, descriptive study, with an analytical comparison of ruptured versus non-ruptured omphaloceles and of deceased versus surviving patients.
The study was carried out in the Department of Paediatric Surgery of the National Hospital Amirou Boubacar Diallo, Niamey, Niger, over 19 months, from 1 August 2023 to 28 February 2025. Data were collected for each patient from admission until discharge or in-hospital death.

All consecutive patients admitted with an omphalocele during the study period were included (exhaustive sampling), with no age restriction. A ruptured omphalocele was defined by a non-intact sac, identified by three features: (i) a central, peri-umbilical defect at the cord insertion; (ii) amniotic membrane remnants at the margins of the defect or over the eviscerated viscera; and (iii) insertion of the cord onto the defect rather than onto normal abdominal wall lateral to it. Gastroschisis was excluded on the basis of its typical para-umbilical location (usually to the right of a normally inserted cord), the constant and complete absence of any covering membrane or membrane remnant, and a normally sited cord separate from the defect; where the membrane was entirely absent.

The two grouping variables were sac integrity (ruptured vs non-ruptured) and in-hospital vital status (deceased vs alive). Variables studied were sociodemographic characteristics; maternal and pregnancy history (maternal age [years; and <20 years], gravidity [primigravida = first pregnancy], antenatal consultations, antenatal ultrasound and diagnosis, place and mode of delivery); neonatal characteristics (sex, term [full-term vs premature], birth weight [g; and low birth weight <2,500 g], age at admission, admission delay [hours]); pre-referral management (nasogastric tube, intravenous access, sac protection, medicalised transport); lesion characteristics (Aitken type, defect diameter, membrane integrity, associated malformations); treatment (conservative [Grob] or surgical [Gross, Schuster, mesh]); and outcome (complications, length of stay, mortality). Omphaloceles were classified according to Aitken as type I (<4 cm, without liver herniation) or type II/giant (>5 cm and/or liver herniation).
The defect was measured at the bedside/intra-operatively with a graduated paper tape measure, as the largest diameter of the fascial (aponeurotic) ring, excluding the herniated sac and its contents.

Categorical variables were compared with the Pearson chi-square test, or the Fisher exact test when any expected cell count was below 5. Continuous variables were compared with the Mann–Whitney U test, normality having been assessed with the Shapiro–Wilk test. A two-sided $p<0.05$ was considered significant.

At the time of the study, no formal institutional ethics committee was available at the hospital. The study was conducted in accordance with the principles of the Declaration of Helsinki, and written informed consent was obtained from the patients' parents or legal guardians prior to data collection and publication.

Results

The Eighty patients with omphalocele were recorded, with a hospital frequency of 5.6% (80/1,422). Mean age at admission was 1.76 days (range: 1 h to 20 days; median: 18 h), and most of the patients were females (52,5% [n=42]). The majority of patients' families resided in an urban area (58.75%, n=47). Regarding prenatal care, only 6.25% (n=5) of the patients' mothers attended ≥6 consultations, and 48.75% (n=39) of them did not have an antenatal ultrasound. Six (14.64%) patients had an antenatal diagnosis of omphalocele. The majority of pregnancies were considered full-term, accounting for 92.50% of the patients (n=74). Premature birth occurred in 6.25% (n=5) of the patients. Hospital delivery occurred in 72.50% (n=58) of the patients, with vaginal delivery being the predominant method 86.25% (n=69). Average birth weight was 2,689.61 g (range: 1,200–4,025 g). Infants with low birth weight (weight <2,500 g) comprised 38.75% of the cohort (n=31).

Average maternal age was 24.40 years (range, 16–46 years). The majority of mothers (86.25%, n=69) were aged 18–35 years. Primigravida represented 26.32% of the population (n=24), whereas primiparous mothers accounted for 27.37% (n=25). Second-degree consanguinity was present in 65% of the patients (n=52). A history of traditional medicine use was present in 72.50% of the mothers (n=58).

Average consultation delay was 45.98 h (range, 1–480 h). Consultation delays exceeding 12 h occurred in 55% of the patients (n=44). The Aitken type 2 (giant omphalocele) omphalocele was identified in 55 patients, representing 68.75% of the cohort. Regarding the amniotic membrane, 41.25% (n=33) of the patients presented with ruptured membranes. Mean defect diameter measured using a tape measure was 4.27 cm for ruptured (range: 3–8 cm) omphaloceles and 4.2 cm for non-ruptured (range: 1.5–10 cm). Associated malformations were present in 13.75% (n=11) of the patients, with cardiac malformations being the most frequent (n=6).



Figure 1: Omphalocele in a 1- day-old female neonate with large umbilical defect

Table 1: Comparison of ruptured and non-ruptured omphaloceles according to different variables

Variable	Ruptured (n=33)	Non-Ruptured (n=47)	Test used	*p*-value
Home delivery	7	15	Chi²	0.42
Prematurity	5	3	Fisher	0.26
Low birth weight	15	13	Chi²	0.16
Birth weight, mean (gm)	2534.5	2665.4	Mann–Whitney	6.54×10^{-19}
Primigravida	15	10	Chi²	0.04
Mother age <20 years	14	6	Chi²	0.005
Mother age, mean	22	25.9	Mann–Whitney	6.54×10^{-19}
Associated malformation	0	15	Fisher	0.0002
Mortality	29	12	Chi²	<0.000001

Table 2: Deceased patients with omphalocele compared with those living with omphalocele according to different variables

Variable	Deceased (n=41)	Living (n=39)	Test used	*p*-value
Home delivery	10	12	Chi²	0.70
Prematurity	4	4	Fisher	1.00
Low birth weight	21	7	Chi²	0.0039
Birth weight, mean (gm)	2512.5	2924.3	Mann–Whitney	6.53×10^{-19}
Primigravida	18	7	Chi²	0.0237
Associated malformation	6	9	Chi²	0.50
Non-medicalised transport	24	25	Chi²	0.78
Admission delay, mean (hour)	8.92	49.35	Mann-Whitney	6.53×10^{-19}
Ruptured	29	4	Chi²	< 0.00000001

There were three patients with Beckwith–Wiedemann syndrome and one set of heteropagus twins with omphalocele. No postnatal amniotic membrane rupture was reported in this study. The ruptured omphaloceles presented with a defect in the umbilical region and eviscerated organs that appeared thickened and/or agglutinated, inflamed, and covered by exudate (Figures 1 and 2). The amniotic membrane was either present as remnants or was completely absent.

The ruptured omphaloceles showed significantly lower birth weights ($p=6.54 \times 10^{-19}$), higher primigravida prevalence (p = 0.04), and younger maternal ages ($p=6.54 \times 10^{-19}$) than the nonruptured omphaloceles (Table 1).
No significant difference was observed in the proportion of patients with prematurity and low birth weight between the ruptured and non-ruptured omphaloceles. Pre-referral management was carried out in more than half of the patients (67.50% [n=54]).

Critical initial management gaps included absent nasogastric tubes (97.5%, n=78), delayed intravenous access (76.25%, n=61), and absence of dressing or protective material for the sac



Figure 2: Ruptured omphalocele with residual amniotic membranes insufficient to cover eviscerated bowel loops

(36.25%, n=29). Transport was non-medicalised in 61.25% of the patients (n=49). Conservative treatment (the Grob technique) was employed for all the non-ruptured omphaloceles, representing 60% (n=48) of the total cohort (Figure 3). Among the surgical interventions performed in 26 patients, the Gross technique with primary closure was the most frequently utilised approach, accounting for 15 (18.75%) of the patients.



Figure 3: Conservative treatment according to the Grob's technique A) Newborn on day 1 of life presenting a type II omphalocele B) Appearance on day 41 of life showing membrane epidermisation C) Appearance on day 50 of life -an ongoing epidermisation D) Appearance on day 71 of life showing an almost complete epidermisation

The Schuster technique was applied in 10 (12.5%) patients, and mesh repair was used in one (1.25%). Six deaths before treatment were recorded.

Length of hospital stay was between 1–55 days (mean, 9.81 days). Complications (18.75%, n=15) included enterocutaneous fistula (n=3) and respiratory distress (n=3).

Mortality rate was 51.25% (n=41), mostly due to respiratory failure (n=19) and sepsis (n=13). The most significant factors that contributed to mortality were omphalocele membrane rupture (p=0.00000001) and a prolonged delay in admission (p=6.53 × 10^{-19}) (Table 2). Factors with a probable but less marked effect were primiparity (p= 0.0237) and low birth weight (p=0.0039). Factors with no significant effect were home delivery (p=0.70), prematurity (p=1.00), associated malformations (p=0.50), and non-medicalised transport (p=0.78).

Discussion

In our study, omphaloceles represented 5.6% of hospitalised cases managed in our paediatric surgery unit. According to reports from Sub-Saharan Africa, omphaloceles account for 4–8% of congenital anomalies, with particularly high rates noted in Uganda and Nigeria.[7] These variations may reflect differences in healthcare access, referral systems, and diagnostic capacity across the region. The majority of the patients in our series involved young mothers with poorly supervised or unsupervised pregnancies.[3] Unsupervised antenatal care and delivery, and absence of prenatal diagnosis led to late presentation and complicated clinical conditions.[2,3]

Primigravida mothers accounted for 26.32% of our population, contrasting with findings from Talabi *et al.* and Abdur-Rahman *et al.* who reported higher parity, with 60% at their third birth or higher, and a mean parity of 3.1, respectively.[2,8] Home delivery accounted for 27.5% of our cases, which is higher than the 19.6% and 16% reported in two Nigerian studies.[2,8] Associated malformations were present in 13.75% of the patients, which is lower than the 30–88% range reported by Talabi *et al.* who similarly identified cardiac and gastrointestinal anomalies as predominant.[2]

A significant proportion of the patients had ruptured omphaloceles (41.25%). Notably, this is a higher rate than those reported in the literature, which is between 7–32.1%.[1-3,8] No membrane rupture in the postnatal period was reported. The aspect of the eviscerated organs similar to that found in gastroschisis suggests the occurrence of membrane rupture during the antenatal period. Previous studies have noted that ruptured omphaloceles with absent membranes may mimic gastroschisis, especially in association with genetic syndromes like Turner syndrome.[9,10] In our series, both ruptured omphalocele and younger maternal age co-occurred; we report this as an observation only and draw no conclusion regarding a shared aetiology. Several studies have consistently identified maternal age <20 years as a significant risk factor for gastroschisis.[12-14] We did not identify any clear causes of antenatal membrane rupture, especially due to limited information about perinatal history. For the post-natal membrane rupture, the size and contents of the omphalocele sac are known factors that increase the risk of rupture. Large or "giant" omphaloceles containing the liver are subject to greater mechanical tension, which increases the likelihood of membrane disruption.[1] Although some authors debate the role of delivery mode as a risk for rupture, there is no consensus. Some studies suggest that vaginal delivery may contribute to rupture in large defects, while others find no significant association.[1] Initial management in our context revealed substantial pre-referral gaps, often delaying resuscitation and increasing the risk of complications. Both conservative and surgical approaches were used, depending on the condition of the sac and available resources. Conservative management is often preferred for intact sacs, especially in resource-limited settings, with delayed surgical closure when feasible.[3,6] Immediate surgical repair is generally reserved for ruptured sacs or when conditions permit safe anaesthesia and postoperative care.[2,3,6]

Factors contributing to poor outcomes include delayed admission, low birth weight, inadequate initial management, and primigravida. Sepsis and postoperative respiratory failure, associated anomalies, large defects, and immediate surgical repair are other factors identified in the literature that contribute to poor outcomes.[2-4] We observed a mortality rate (51.25%) significantly higher than those found in the literature (between 22.9–42%).[2-4, 8] In Sub-Saharan countries, sepsis is among the leading causes of death.[2-4]

Conclusion

Omphalocele was frequent in our setting (5.6% of admissions) and carried a high mortality (51.25%). In this cohort, a ruptured sac, low birth weight, delayed admission and primigravidity were each associated with mortality. The high proportion of ruptured omphaloceles and the factors underlying rupture were not elucidated by this study and warrant further investigation.

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