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The risk of midgut volvulus in patients with abdominal wall defects: A multi-institutional study

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ABSTRACT

Background: The management of malrotation in patients with congenital abdominal wall defects has varied among surgeons. We were interested in investigating the risk of midgut volvulus in patients with gastroschisis and omphalocele to help determine if these patients may benefit from undergoing a Ladd procedure.
Methods: A retrospective chart review was performed for all patients managed at three institutions born between 1/1/2000 and 12/31/2008 with a diagnosis of gastroschisis or omphalocele. Patient charts were reviewed through 12/31/2012 for occurrence of midgut volvulus or need for second laparotomy.
Results: Of the 414 patients identified with abdominal wall defects, 299 patients (72%) had gastroschisis, and 115 patients (28%) had omphalocele. The mean gestational age at birth was 36.1 ± 2.3 weeks, and the mean birth weight was 2.57 ± 0.7 kg. There were a total of 8 (1.9%) cases of midgut volvulus: 3 (1.0%) patients with gastroschisis compared to 5 patients (4.4%) with omphalocele ($p = 0.04$).
Conclusions: Patients with omphalocele have a greater risk of developing midgut volvulus, and a Ladd procedure should be considered during definitive repair to mitigate these risks.
Level of evidence: III; retrospective comparative study

The normal rotation of the intestine follows precise embryologic events that result in a fixed intestinal configuration with the duodenojejunal junction in the left upper quadrant and the cecum in the right lower quadrant [1]. Normal intestinal configuration allows for the midgut to be anchored by a wide mesenteric base. Patients with abdominal wall defects have a disruption in embryologic rotational events, which results in malrotation of the intestines with potential for a narrow mesenteric base and midgut volvulus. Midgut volvulus is a devastating event associated with significant morbidity and mortality. The Ladd procedure is the procedure of choice for correction of malrotation.

Performing a Ladd procedure in patients with abdominal wall defects is variable among surgeons [2–5]. This variability is a result of the belief that adhesions from repair of the abdominal wall defect should be sufficient to provide bowel fixation and prevent midgut volvulus. Additionally, manipulation of friable bowel may pose risk to the patient [2,3]. However, the ability of adhesions to provide bowel fixation remains uncertain. There is little evidence regarding the incidence of midgut volvulus in patients with abdominal wall defects. Furthermore, gastroschisis patients may have more protective adhesions because of inflammation and increased handling. However, this is not true in the case of omphalocele and may predispose these patients to midgut volvulus. In a single-institution study examining midgut volvulus in subjects with abdominal wall defects, 3% of those with omphaloceles developed midgut volvulus. Conversely, none of those with gastroschisis developed midgut volvulus [6]. However, because of the relatively low incidence of abdominal wall defects, we sought to expand on these data by performing a multi-institutional study. The aim of this study was to examine the occurrence of midgut volvulus in patients with abdominal wall defects and identify a group who may be at a greater risk.

1. Methods

Data were collected from three participating institutions: Children's Hospital of Wisconsin (Milwaukee, WI), Children's Mercy Hospital (Kansas City, MO), and The Children's University Hospital (Dublin, Ireland). After institutional review board approval, charts of all patients with an ICD-9 diagnosis code for gastroschisis (756.73) and omphalocele (756.72) were retrospectively reviewed. Patients were included if their birth dates were between the ranges of 1/1/2000 through 12/31/2008. The patients' charts were reviewed including all emergency room visits,

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hospital admissions, and outpatient clinic appointments, through 12/31/2012 to assess if these patients developed midgut volvulus. Patients were excluded if they had an abdominal wall defect that was not gastroschisis or omphalocele, if they died prior to surgical repair of their defect, or if their primary surgical repair occurred at an outside facility. Small omphalocele was defined as <4 cm defect, and giant omphalocele was defined as >4 cm defect or defect containing the liver [7]. Midgut volvulus was defined as twisting of the midgut around its mesentery in the setting of malrotated bowel without predisposing abnormalities including adhesions, congenital band, meconium ileus, or mass.

Electronic medical records were reviewed for patient demographics, outcomes including details of surgical repair (primary vs delayed, need for patch, Ladd procedure completed on final repair), and surgical complications requiring a second unplanned laparotomy (midgut volvulus, adhesive bowel obstruction, internal hernia, abdominal compartment syndrome). Primary outcome was defined as the occurrence of midgut volvulus. Secondary outcomes were need for unplanned laparotomy and death.

Data were collected and stored in the Research Electronic Data Capture (REDCap™) system. Patient factors and outcomes were compared between the gastroschisis and omphalocele groups using the chi-square test and Fisher's exact test for categorical variables and the Wilcoxon rank-sum test for continuous variables (* in Tables 1 and 4 indicate Fisher's exact test). *P* values ≤0.05 were considered statistically significant. All statistical analyses were performed using SAS 9.3 software (SAS Institute, Cary, NC).

2. Results

During the defined study period, there were a total of 414 patients with an abdominal wall defect. Of these, 299 (72.2%) patients had a diagnosis of gastroschisis, while 115 (27.8%) had a diagnosis of omphalocele. Of those with omphaloceles, 54 (50.0%) had a small omphalocele, while 36 (31.3%) had a giant omphalocele. There were 25 (21.7%) with an unknown size of the omphalocele. Neonatal characteristics are summarized in Table 1. The mean gestational age at delivery was 36.1 ± 2.3 weeks. Those with gastroschisis were delivered earlier than those with an omphalocele (36.0 ± 1.9 vs 36.5 ± 2.9; $p = 0.001$). The mean birth weight was 2.57 ± 0.7 kg in gastroschisis patients having a statistically significantly lower birth weight (2.49 ± 0.55 vs 2.90 ± 0.9; $p = <0.0001$). Furthermore, the mortality rate was statistically greater in those with an omphalocele (4.0% vs 11.3%; $p = 0.005$) (Table 2).

In the study cohort, there were a total of 8/414 (1.9%) subjects who developed midgut volvulus (Table 3). The operation performed was reduction of volvulus and bowel resection as indicated. There was a statistically significant difference in the occurrence of midgut volvulus between those with gastroschsis and those with omphalocele. In omphalocele patients, 5/115 (4.4%) developed midgut volvulus, whereas 3/299 (1.0%) of subjects with gastroschisis developed midgut volvulus ($p = 0.04$). Of the omphalocele patients who developed midgut volvulus, 2/5 (40%) were categorized as small and 3/5 (60%) had giant omphaloceles. Furthermore, the median age that midgut volvulus occurred was 1098 days (range, 21–2285 days). However, the occurrence of midgut volvulus tended to cluster at two time points. Four of the patients had midgut volvulus within or just after the first year of age. The other four developed midgut volvulus around 5–6 years of age. Three of those who developed midgut volvulus required a bowel resection. However, volvulus was not a cause of short bowel syndrome for any patients. Additionally, volvulus was not a cause of mortality in this cohort.

Only 8/414 (5.1%) of the study cohort underwent a prophylactic Ladd procedure at some point during repair of the abdominal wall defect. There was a difference in the rates of Ladd procedures between the two groups. Of those with gastroschisis, 3/299 (1.0%) had a Ladd procedure, whereas 5/115 (4.3%) of those with omphaloceles underwent a Ladd procedure. All Ladd procedures were performed on an elective basis. The decision to perform a Ladd procedure seemed to be solely based on surgeon preference. All Ladd procedures were performed at definitive closure. Furthermore, a single institution in the study performed all 8 of the Ladd procedures (Children's Hospital of Wisconsin). None of the patients who underwent a Ladd procedure later developed midgut volvulus. Additionally, none of those who underwent a Ladd procedure required a laparotomy secondary to an adhesive bowel obstruction.

A significant portion of the study cohort required a laparotomy for nonvolvulus-related complications (Table 4). A total of 57/414 (13.7%) required a laparotomy for any complication with an equal rate between gastroschisis and omphaloceles (14.0% vs 13.0%; $p = 0.874$). The most common complication among the entire cohort requiring a laparotomy was adhesive bowel obstruction occurring in 25/414 (6.0%) of the total population. There was not a difference in the rate of adhesive bowel obstruction between gastroschisis and omphalocele (6.6% vs 4.4%; $p = 0.491$). However, the rate of adhesive bowel obstruction was equal to that of midgut volvulus in patients with omphalocele (4.4% vs 4.4%). This was not the case for patients with gastroschisis, with bowel obstruction being 6 times more common (6.6% vs 1.0%).

3. Discussion

Midgut volvulus is a catastrophic complication of malrotation, which is inherent in patients with abdominal wall defects. Although the association between congenital abdominal wall defects and intestinal rotational anomalies is well established, performing a Ladd procedure is not standard of care [2–5]. This is because of operative challenges and perhaps the belief that adhesions are sufficient enough to provide bowel fixation [2,3].

Table 1
Demographics.

Patient demographics	Total (n = 414)	Gastroschisis (n = 299)	Omphalocele (n = 115)	*p*-value
Sex				0.3416
Male	204 (49.3%)	143 (47.8%)	61 (53.0%)	
Female	210 (50.7%)	156 (52.2%)	54 (47.0%)	
Gestational age	36.1 ± 2.3	36.0 ± 1.9	36.5 ± 2.9	0.0013
Birth weight	2.57 ± 0.7	2.49 ± 0.55	2.90 ± 0.9	<0.0001
Ethnicity				0.0773
Caucasian	278 (67.1%)	195 (65.2%)	83 (72.2%)	
African American	48 (11.6%)	32 (10.7%)	16 (13.9%)	
Hispanic	46 (11.1%)	41 (13.7%)	5 (4.4%)	
Asian	11 (2.7%)	9 (3.0%)	2 (1.7%)	
Other/unknown	31 (7.5%)	22 (7.4%)	9 (7.8%)	
Midgut volvulus	8 (1.9%)	3 (1.0%)	5 (4.4%)	0.0408*
Neonatal death	25 (6.0%)	12 (4.0%)	13 (11.3%)	0.0053
Ladd procedure for incidental malrotation	8 (1.9%)	3 (1.0%)	5 (4.3%)	0.0408