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Volvulus without malposition—a single-center experience

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ABSTRACT

Background: This is a single-center case series about the rare condition of volvulus without malposition and/or malrotation (VWM) in preterm babies. We focus on diagnostic difficulties, and our results should help to distinguish VWM as a distinct entity different from classical volvulus and segmental volvulus.

Materials and methods: Medical chart review of infants with VWM from 2003–2012 was used.

Results: A total of 15 patients were identified. All of them had volvulus in the absence of intestinal malposition or other associated intestinal pathologies. All patients were born prematurely. Emergency laparotomy was necessary in all 15 patients. Two groups were identified. Group 1 includes four patients with typical signs of meconium obstruction of prematurity (MOP). Small bowel resection was only necessary in one of these four patients, all survived without residual intestinal lesions. Group 2 consists of 11 patients without signs of MOP—small bowel resection and temporary enterostomy were necessary in all these children. Four patients presented with pneumatosis intestinalis on the abdominal plain film, suggesting necrotizing enterocolitis. Although two infants died, the survivors showed complete recovery.

Conclusions: VWM is a distinct disease of prematurity. When associated with MOP, VWM has a favorable outcome of treatment. In contrast, VWM occurring in the absence of signs of meconium obstruction requires small bowel resection. VWM primarily affects the top of the midgut (ileum). Because of absent malposition, presentation of VWM may be uncharacteristic. Pneumatosis intestinalis in advanced VWM may lead to diagnostic difficulties and a delay in treatment.

1. Introduction

An increasing rate of preterm babies can be treated with a favorable outcome concerning their intrinsic problems of prematurity. This has confronted neonatologists and pediatric surgeons with a number of gastrointestinal diseases exclusively found in this period of life. Although necrotizing enterocolitis (NEC), spontaneous intestinal perforation (SIP), and meconium obstruction of prematurity (MOP) have been widely discussed, the phenomenon of volvulus without malposition (VWM) represents a commonly underestimated or even neglected diagnosis in differential diagnosis of the acute abdomen in preterm babies. We define VWM as volvulus in the absence of an intestinal congenital anomaly of rotation of the midgut (malrotation).

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We report on a series of 15 children with VWM. Particularly, we highlight the difficulties in establishing a correct diagnosis without delay, which may possibly result in severe consequences in the affected patients. Awareness of this rare intestinal emergency is helpful in the sometimes challenging workup of gastrointestinal diseases of preterm babies.

Additionally to our results, we propose an accurate differentiation between "classical volvulus" (associated with malposition), "segmental volvulus" (causative anatomic anomaly), and "VWM". On careful consideration, these three pathologic entities differ in terms of clinical and radiological presentation and outcome of treatment.

2. Methods

Patients with VWM were identified in our pediatric surgical database from 2003–2012, and medical records were reviewed. Infants with segmental volvulus associated with mesenteric defects or intestinal cysts or tumors were excluded. Medical records were reviewed for demographic data, clinical and radiographic findings, timing of diagnosis and intervention, complications and outcome of treatment.

3. Results

We identified 15 infants with VWM of the intestine. These 15 infants were divided into two groups based on intraoperative findings as follows: group 1 includes 4 patients with typical signs of MOP whereas group 2 consists of 11 patients without any signs of MOP. Patient characteristics are shown in Table 1. Gestational age ranged from 24–33 wk (median age 26 wk). The majority of these infants were delivered by cesarean section (12/15). Respiratory support (continuous positive airway pressure and/or mechanical ventilation) was necessary in 10 neonates immediately postpartum. Oral feedings have been initiated in the first days of life, starting with few milliliters of glucose and electrolyte solution advancing to preterm formula feeds.

All patients presented with severe abdominal distention. In five patients, a livid discoloration of the abdominal wall was visible. Three infants were passing bloody stool. None of the patients presented with bilious and/or green vomiting, suggestive of high intestinal obstruction—the leading symptom of classical volvulus.

Preoperative serum lactic acid, c-reactive protein, and white blood cell count showed a wide range (Table 2). Laboratory findings assessed at the day of surgery did not correlate with the extent of bowel damage.

Repeated abdominal plain films and abdominal ultrasound examinations were done in all cases. The leading sign was significant dilation of bowel loops and absence of air in the pelvic region on plain abdominal x-ray, suggesting intestinal obstruction (7/15). Two patients had a gasless abdominal x-ray. Four patients presented with intestinal pneumatosis in abdominal plain film and abdominal ultrasound. In all other patients, abdominal ultrasound examination revealed nonspecific findings such as reduced peristalsis, ascites, or dilated bowel loops. A "whirl-sign," which is pathognomonic for volvulus could not be found—although the radiologists paid special attention to the position of the superior mesenteric vein and artery both central and peripheral.

Transverse right upper quadrant laparotomy was performed in all 15 children. In 13 of 15 cases, the preoperative diagnosis indicating emergency laparotomy was volvulus. A suspected diagnosis of advanced NEC due to intestinal pneumatosis finally led to laparotomy in two infants. The incorrect preoperative diagnosis of NEC resulted in a delay of definite treatment (laparotomy) in one patient negatively affecting the outcome of treatment (Table 1, patient 4, group 2).

In four children (group 1), laparotomy revealed the terminal ileum to be filled with inspissated meconium leading to small bowel obstruction—a condition described as MOP. Intestinal obstruction finally led to a clockwise twist of ileal loops. In 11 patients (group 2), no signs of MOP were found at laparotomy. Clockwise volvulus including the top of the midgut loop (ileum) resulted in necrotic small-bowel segments in all these patients.

Infants in group 1 were younger at diagnosis and had a longer duration of symptoms preceding abdominal surgery than infants of group 2. There were more consultations by a surgeon in group 1 before laparotomy. Table 3 presents relevant differences between group 1 and group 2.

In group 1 (VWM and MOP), volvulus led to small bowel necrosis necessitating resection in only one infant. In the other three patients, the intestines were viable after untwisting and removal of the inspissated meconium. The four infants of group 1 fully recovered.

In all 11 children of group 2, resection of at least 25 cm ileum and ileostomy were necessary. Nine neonates showed complete recovery after intestinal reconstruction, and none of the survivors developed short bowel syndrome. Of the two deaths in this group, one died from sepsis after advanced intestinal necrosis, whereas the other one died of severe extraintestinal complications of prematurity.

Gestational age and weight at the time of laparotomy did not correlate with an adverse outcome. Intestinal reconstruction was successfully performed at an average of 67 d (11–149) after the initial procedure.

4. Discussion

VWM is one of the diseases associated with prematurity that is emerging because of increased survival rates of preterm babies. Although gastrointestinal problems are frequently found in preterm babies, the phenomenon of VWM has not received much attention in the past.

Classical volvulus is a well-known intestinal emergency predominantly occurring in the neonatal period. Because of malposition, the intestinal convolute hangs freely like a grape in the abdominal cavity prone to twisting. Classical volvulus is therefore a clockwise rotation of the entire small bowel and the right hemicolon around the mesenteric axis (superior mesenteric artery) on the basis of a congenital malposition of the intestine. Classical volvulus is extremely rare in preterm babies [1]. In most of the cases of intrauterine volvulus, no malposition or anatomic abnormality is found leading to classification as VWM [2].