



## Malrotation of the Intestine

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Malrotation of the intestinal tract is a product of a well defined aberrant embryology. Because the consequences of malrotation associated with a midgut volvulus may be catastrophic, an understanding of the anatomy, diagnostic criteria, and appropriate therapy for this putative emergency illness is imperative. This report summarizes a recent 18-month experience with this diagnosis and contrasts this experience with that in the published literature. More than half (14/22) of the patients presented during the first month of life, and all had vomiting, which in most cases was bilious. The barium upper gastrointestinal series was the preferred diagnostic study, being both sensitive (18/19, 95%) and accurate (18/21, 86%). In this series two-thirds of the patients presented with volvulus (15/22, 68%) of whom five had ischemic intestine requiring resection. One of these children died of overwhelming sepsis. A Ladd procedure was the preferred treatment, which as defined by us includes evisceration and inspection of the mesenteric root, counterclockwise derotation of a midgut volvulus, lysis of Ladd's bands with straightening of the duodenum along the right abdominal gutter, inversion-ligation appendectomy, and placement of the cecum into the left lower quadrant. A high index of suspicion in the neonate with vomiting, rapid diagnosis, and appropriate operative therapy results in a predictable favorable outcome for children with intestinal malrotation.

Malrotation may be thought of as the sentinel case that has distinguished the field of pediatric surgery. It is based on a defined embryology; it requires a high index of suspicion with excellent clinical judgment to establish its diagnosis; and its treatment is based on the classic description by Ladd [1], acknowledged by many to be the founding father of the field of this surgical subspecialty.

This report reviews the precise embryology of intestinal rotation and malrotation, the clinical features of malrotation with and without volvulus, the criteria for diagnosis, and finally the techniques of treatment. The basis for this review is an 18-month series from this medical center that complements data published in the world's literature.

### Materials, Methods, and Results

#### Patient Review

From January 1990 to June 1991, 22 patients underwent operation in this hospital for malrotation with and without volvulus

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(Table 1). There were 12 males and 10 females in the series. Nineteen were white and three were black. The age at operation ranged from 1 to 1095 days (mean 124.7 days, median 20 days) with 6 being under 1 week of age, 14 being less than 3 weeks of age, and 19 being under 6 months of age.

#### Patient Presentation and Diagnosis

All patients presented with intermittent acute onset or recurrent vomiting, which was bilious in 17 (77%). The physical examination was recorded as being normal in 11 (50%), distension was present in 8 (36%), and the remaining 3 (14%) had tenderness and signs of peritoneal irritation (Table 1).

Radiographic upper gastrointestinal contrast studies were done in 19 patients and the diagnosis was accurate in 18. One child was inappropriately thought to have gastroesophageal reflux without malrotation. This 95% sensitivity (18/19) of the barium meal contrasts with the accuracy of the study, 86% (18/21), as during this time interval two additional patients labeled as "malrotation" radiographically were found to have normal position of the ligament of Treitz at laparotomy. The only barium enema examination done in this patient series was thought to be normal.

#### Patient Treatment and Outcome

Of the 22 patients, 15 (68%) had volvulus; and of these 15, 5 required resection. All patients underwent a Ladd procedure, which included derotation of the volvulus, lysis of Ladd's bands, straightening of the duodenum in the right flank, and placement of the cecum into the left lower quadrant after inversion-ligation appendectomy. The one death (4.5%) in this series occurred in a 2-week-old who presented with an acute abdomen, volvulus, and sepsis. The patient died on the second postoperative day of overwhelming infection.

### Discussion

#### Embryology

The embryology of intestinal rotation was described by Mall in 1898 [2]. Twenty-five years later, Dott was the first to clearly

**Table 1.** Malrotation with or without midgut volvulus operated at the Children's Hospital Medical Center of Cincinnati, January 1990 to June 1991.

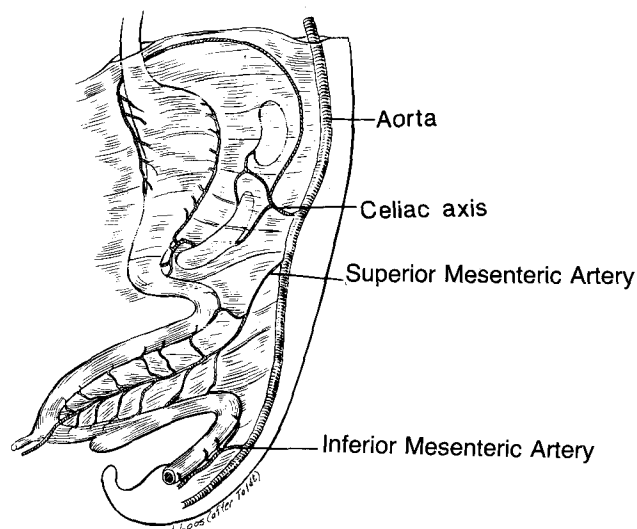
Pt.	Age	Race/sex	Symptoms	Abdom. exam	Ugl	Lgl	Ladds	Resection	Volvulus
1	1 day	W/M	bV	dist	mal + obst	ND	yes	yes	yes
2	1 day	W/F	V + dist	dist	ND	ND	yes	no	no
3	2 days	W/M	bV	dist	mal	nml	yes	yes	yes
4	3 days	W/M	bV	nml	mal	ND	yes	no	yes
5	5 days	W/M	bV	dist	mal	ND	yes	no	yes
6	7 days	W/F	bV	tender	mal	ND	yes	yes	yes
7	11 days	W/F	bV	dist	mal + obst	ND	yes	no	yes
8	12 days	W/M	bV	nml	mal + obst	ND	yes	no	no
9	14 days	W/M	V	acute	ND	ND	yes	yes	yes
10	14 days	B/F	bV	acute	ND	ND	yes	yes	yes
11	19 days	W/F	bV	dist	mal	ND	yes	no	yes
12	21 days	W/F	bV + FTT	nml	mal	ND	yes	no	yes
13	21 days	W/M	bV	nml	mal	ND	yes	no	no
14	21 days	W/M	V	nml	mal + obst	ND	yes	no	no
15	7 weeks	W/M	bV	nml	mal + obst	ND	yes	no	yes
16	2 months	W/F	bV	nml	mal	ND	yes	no	no
17	3 months	W/F	bV	dist	mal	ND	yes	no	no
18	4 months	W/F	bV	nml	mal	ND	yes	no	no
19	6 months	B/M	bV	nml	mal	ND	yes	no	yes
20	15 months	B/M	bV	nml	mal	ND	yes	no	yes
21	1.5 years	W/F	V	nml	GER	ND	yes	no	yes
22	3 years	W/M	V + pain	dist	mal	ND	yes	no	yes

W: white; M: male; B: black; F: female; b: bilious; nml: normal; mal: malrotation; V: vomiting; dist: distended; FTT: failure to thrive.

There were 12 boys and 10 girls. They all presented with intermittent, acute or recurrent vomiting, and it was bilious in 77% of the cases. Physical examination was normal in 50% of patients, and distension was present in 32%. Sixty-eight percent of the patients had a volvulus, and 23% required resection and primary anastomosis. Patient 9 died on postoperative day 2 of overwhelming sepsis.

correlate the observations of the embryologist and the associated clinical problems [3]. In 1936 Ladd wrote the classic article on the treatment of the condition, describing 21 cases in which treatment was based on an understanding of the underlying embryology [1].

In the 5-mm embryo, at about the 4th week of embryologic development, the midgut extends from the anterior to the posterior intestinal portal and communicates widely with the yolk sac by way of the vitelline duct. The development of the midgut is characterized by rapid elongation resulting in the formation of the primary intestinal loop (Fig. 1). The cephalic end represents the prearterial, or duodenojejunal, loop. The caudal limb represents the postarterial, or ileocecal, loop, which extends from the distal ileum to the proximal two-thirds of the transverse colon. Physiologic umbilical herniation occurs during the 6th week of development. Coincident with the growth in length, the primitive intestinal loop rotates through an arch of 270 degrees around an axis formed by the superior mesenteric artery (SMA) in a *counterclockwise* direction (Fig. 2). It results in the normal location of the second portion of the duodenum to the right of the SMA, the third portion of the duodenum posterior to the SMA, and the duodenojejunal junction to the left and posterior to the SMA. At approximately the end of the 3rd month the herniated intestinal loop begins to return to the abdominal cavity. During the 4th and 5th months of development, the mesentery becomes firmly attached to the posterior abdominal wall in a diagonal plane extending from the cecum in the right lower quadrant to the duodenojejunal junction in the left upper quadrant. The junction of the cranial and caudal limbs in the adult can be recognized only if a portion of the vitelline duct persists as a Meckel's diverticulum.



**Fig. 1.** Primitive midgut rapidly elongates and forms the primary intestinal loop supplied by the superior mesenteric artery.

Malrotation of the midgut occurs when the normal rotational process and fixation of the intestine fail to take place. The basic embryologic development of the intestine permits classification of these anomalies into three stages as described by Dott [3], Frazer and Robbins [4], Snyder and Chaffin [5], and Kiesewetter and Smith [6].

*Stage one: herniation.* This stage takes place during the first 10 weeks of development when the midgut protrudes through

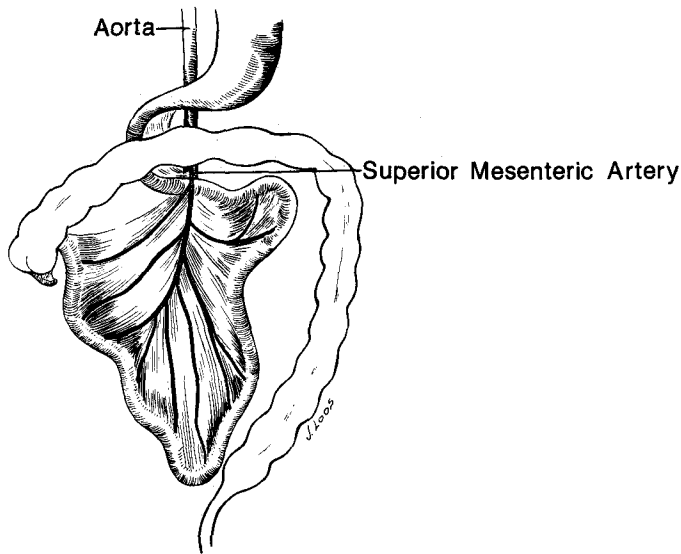


Fig. 2. At about the 11th week of embryologic development, the midgut loop rotates 270 degrees on an axis formed by the superior mesenteric artery.

the vitelline sac. Failure of the rotation to go beyond this stage results in an omphalocele.

*Stage two: return to the abdomen.* This stage occurs between the 10th and 11th weeks of development. The midgut retracts from the umbilical cord and returns to the abdominal cavity. Normally, the prearterial or duodenojejunal limb returns first and continues its *counterclockwise* 270-degree rotation around the SMA axis. Failure of this event to occur leads to a spectrum of abnormalities, including nonrotation, incomplete rotation, paraduodenal hernia, and reversed rotation. Nonrotation occurs when the duodenum descends straight down to the right side of the abdomen, and the colon lies on the left. This condition is usually asymptomatic. Incomplete rotation, also known as malrotation, causes duodenal obstruction either by bands or volvulus. The duodenal loop lacks 90 degrees of its normal 270-degree rotation, and the cecocolic loop lacks 180 degrees of its normal rotation. Internal paraduodenal hernia is rare.

Reversed rotation may occur when either the postarterial limb returns to the abdominal cavity first and rotates in a counterclockwise direction for 180 degrees or when the primitive loop rotates 90 degrees in a clockwise direction. With such an abnormality, the transverse colon passes behind the duodenum; this problem accounts for about 4% of cases [7-9], and it is associated with partial mesenteric arterial, venous, and lymphatic obstruction. Absence of the superior mesenteric venous drainage has been reported with this anomaly [10].

*Stage three: fixation.* Fixation takes place from the 12th week until birth. Its failure can result in a mobile cecum, a subhepatic cecum, or a retrocecal appendix.

### Clinical Presentation

The incidence of malrotation or nonrotation of the midgut is approximately 1 in 500 live births [11]. Two-thirds (64% in our series, 14/22) present during the newborn period. Males pre-

dominate 2:1 in this age group (8 males, 6 females in our series). Approximately 20% (14%, 3/22 in our series) present after 1 year of age, and here the male predominance disappears.

Anomalies of rotation and fixation of the gastrointestinal tract are frequently associated with abdominal wall defects and diaphragmatic hernia. Nonrotation is a part of the developmental defect in these entities. Associated gastrointestinal anomalies are found in up to 62% of the reported cases [12], with intestinal atresias being the most common [12-17]. Volvulus occurring in utero may produce varying degrees of intestinal atresia. In addition, intraluminal duodenal membranes may be present; and unless specifically excluded by passage of an intraluminal tube, they may be overlooked during a Ladd's procedure. Other associated problems include Meckel's diverticulum, intussusception, Hirschsprung's disease, gastroesophageal reflux, gastric volvulus, persistent cloaca, and anomalies of the extrahepatic biliary system [11-28].

Most patients present with emesis. Bilious vomiting is the hallmark of malrotation and midgut volvulus, especially in the newborn. In our series, all patients presented with vomiting, and it was bilious in 77%. In the older child, other common presenting symptoms may include intermittent vomiting, failure to thrive, and recurrent abdominal pain. Each was seen in our series. Volvulus of the midgut occurs less frequently, approximately 14%, when compared to 68% in neonates with malrotation. In our series 71% (10/14) under 3 weeks of age had volvulus, whereas 63% (5/8) older than that age had volvulus.

Strikingly, physical examination may reveal a totally normal abdominal picture, a feature recorded in 50% (11/22) of our patients. Distension without tenderness is the next most common sign and occurred in an additional 7 (32%) of our cases. Only those in whom the volvulus has progressed to infarction demonstrate the signs and symptoms of shock and peritonitis. The onset of symptoms may be acute, chronic, or recurrent. A high index of suspicion is essential when evaluating the child with signs and symptoms of small bowel obstruction associated with bilious vomiting.

### Diagnosis

An expeditious workup in conjunction with resuscitation measures should be instituted. Plain films of the abdomen may reveal proximal gastric or duodenal distension, with or without distal intraluminal air. The duodenal triangle has been reported as a plain film sign of midgut malrotation and volvulus in the neonate [29]. Triangular gas shadows in the right upper quadrant are produced as a consequence of the liver edge overlying the air-filled duodenum, and these shadows may be best demonstrated by an upright film. Volvulus cannot be excluded by clinical findings alone; nor is it excluded by a normal abdominal plain radiograph.

In patients where malrotation and volvulus are being considered and no evidence of peritonitis exists, one should proceed with contrast studies. A barium meal is considered the diagnostic study of choice, looking for an abnormal position of the duodenojejunal junction usually to the right of the midline but occasionally to the left; obstruction of the second or third portion of the duodenum with a spiral "beak or corkscrew" appearance; and a proximal jejunum located on the right side of the abdomen [12] (Fig. 3). However, the physician must be

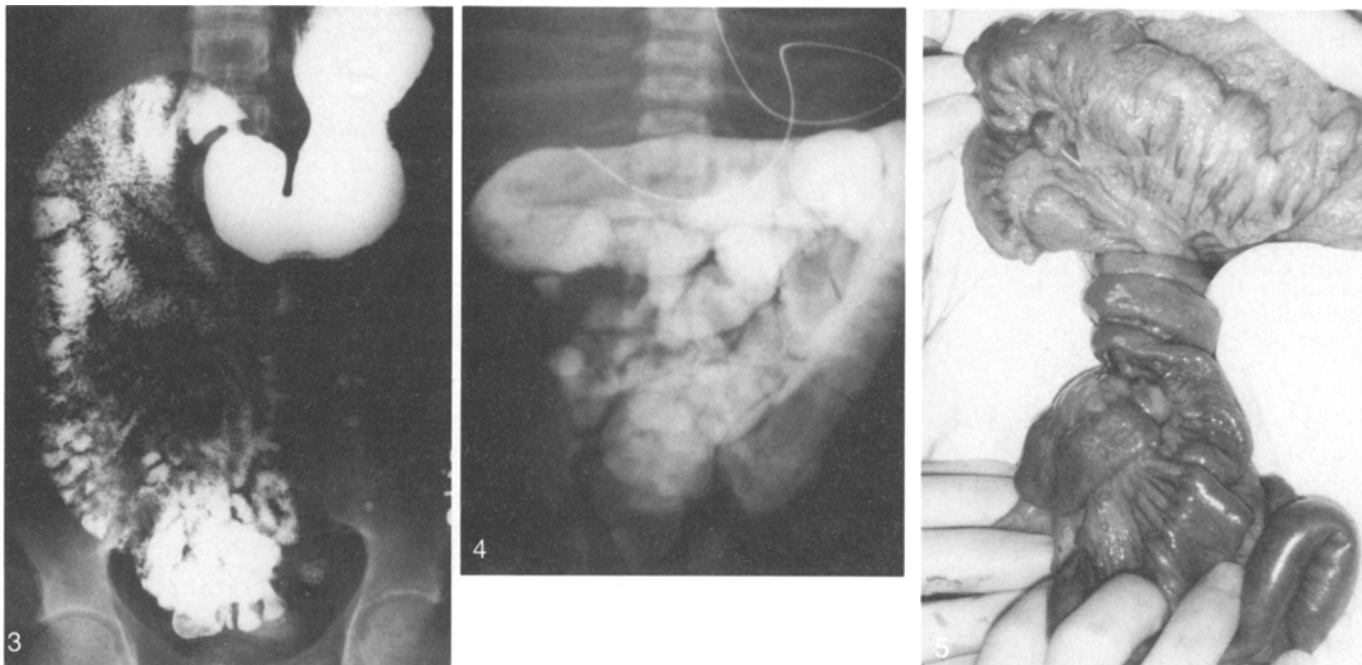


Fig. 3. Barium meal showing the duodenojejunal junction to the right of the spine and proximal jejunal loops in the right side of the abdomen.

Fig. 4. Barium enema demonstrates malrotation of the colon, with the cecum in an abnormal position high and to the left of the midline.

Fig. 5. Intraoperative photograph of a midgut volvulus.

aware that the normal duodenojejunal junction can be readily displaced in neonates; therefore a mobile duodenum discovered on fluoroscopic examination or by positioning a transpyloric feeding tube should not be considered indicative of malrotation in the asymptomatic infant [30].

To exclude malrotation conclusively by radiographic technique, the duodenojejunal junction (ligament of Treitz) must be located to the left of the spine at the level of the duodenal bulb in a position halfway between the lesser and greater stomach curvature.

In the patient with equivocal barium swallow findings, further diagnostic information can be obtained by a barium enema (Table 2). An enema may show the cecum and proximal colon in the left flank or they may cross to the right and then angle back to point to the left hypochondrium (Fig. 4). Malrotation is associated with a normally positioned cecum in 5% to 20% of cases [6, 30, 31]. Sonography, computed tomography, and magnetic resonance imaging have been added by some centers to the diagnostic armamentarium of malrotation when other diagnostic modalities fail to establish the diagnosis [32–34]. Such studies may demonstrate the abnormal orientation of the mesenteric vessels. Normally, the superior mesenteric vein (SMV) lies ventral and to the right of the SMA. In patients with malrotation, the SMA lies immediately anterior to the SMV. Caution should be used when using the SMV rotation sign in patients with hepatomegaly, aortic aneurysm, or marked spinal curvature with abdominal asymmetry, as the SMV may appear to lie on the left ventral aspect of the SMA even when malrotation is not present, leading to a false-positive diagnosis. Angiography has been reported to be helpful when other

Table 2. Accuracy of diagnostic contrast studies of malrotation with or without volvulus.

Author	Barium meal <sup>a,b</sup>	Barium enema <sup>a</sup>
Snyder & Chaffin [5]	10/11 (91%)	Abnormal 9/9 (100%)
Stewart et al. [11]	16/16 (100%)	Abnormal 16/17 (94%) Normal 1/17 (6%)
Krasna et al. [38]	2/2 (100%)	Abnormal 2/2 (100%)
Potts et al. [29]	12/12 (100%)	Abnormal 2/4 (50%) Normal 2/4 (50%)
Yanez & Spitz [20]	31/35 (89%)	Abnormal 13/17 (86%) Normal 2/17 (12%)
Millar et al. [31]	74/78 (95%)	Abnormal 24/29 (83%) Normal 5/29 (17%)
Powell et al. [21]	29/29 (100%)	Abnormal 24/29 (83%) Normal 5/29 (17%)
Welch et al. [41]	14/21 (67%)	Abnormal 13/16 (81%)

<sup>a</sup>The numerator denotes the number of abnormal studies. The denominator denotes the total number of studies.

<sup>b</sup>All abnormal.

diagnostic studies are inconclusive. Arterial changes of a “barber pole sign,” due to a whirling of the SMA and its branches and a tortuous, dilated SMV have been described. Complete occlusion of the SMA and SMV with development of collaterals are angiographic indications of vascular compromise in chronic volvulus secondary to malrotation [35]. Suffice it to say that these time-consuming studies are not applicable in an acutely ill infant suspected to have malrotation and volvulus where laparotomy itself may prove to be the most prudent diagnostic study.

**Table 3.** Summary of midgut malrotation of previously reported cases.

Author	No. of pts.	Sex (M:F)	Incidence (%)		Emesis	Volvulus	Resection	Mortality
			Age					
			< 1 mo	> 1 mo				
Snyder & Chaffin [5]	40	26:16	88	12	100	80	*	23
Kiesewetter et al. [6]	44	27:17	80	20	86	45	*	20
Wang & Welch [7]	6	0:6	0	100	*	50	0	0
Stewart et al. [11]	159	*	64	36	*	48	15	18
Krasna et al. [38]	5	4:1	100	0	80	100	80	20
Andrassy & Mahour [18]	74	51:23	53	47	100	*	*	4
Filston & Kirks [12]	34	*	71	29	*	50	*	15
Brandt et al. [19]	16	*	0	100	44	6	*	6
Potts et al. [29]	45	25:20	82	18	100	96	*	24
Yanez et al. [20]	37	19:18	0	100	41	14	*	5
Young et al. [13]	3	1:2	33	67	66	0	*	0
Katz & Spitz [30]	35	20:15	*	*	57	29	*	*
Millar et al. [31]	137	2:1	62	38	60	63	15	19
Kumar et al. [25]	40	*	0	100	100	*	*	2.5
Powell et al. [21]	70	*	39	*	66	44	21	10
Welch et al. [41]	34	20:14	100	0	100	71	18	6

\*: not mentioned.

### Operative Management

If volvulus is suspected, immediate and aggressive resuscitation must be achieved before urgent operation. A supraumbilical right abdominal transverse muscle cutting incision is made, and the entire small and large bowel is eviscerated. The root of the mesentery is inspected and, if present, the volvulus is untwisted in a *counterclockwise* fashion (Fig. 5). The viability of the bowel is then assessed. Warm lap pads may be used to cover the bowel. The right upper quadrant Ladd's bands stretching across the duodenum are next incised, and the duodenum is straightened so it lies comfortably in the right flank. The pedicle of the mesentery is dissected to broaden its base. The small bowel is inspected, and its intraluminal contents are milked distally to assess intestinal luminal patency. It is prudent to pass a balloon catheter through the duodenum to exclude a partially obstructive intraluminal web or membrane. The mobilized cecum is placed into the left lower quadrant, and the rest of the small bowel is placed on the right side of the abdomen. We routinely perform an inversion-ligation appendectomy prior to returning the cecum to the left lower quadrant. We have not suture-fixed the cecum into this new position, though prophylactic fixation of the intestine in the corrected malrotated position has been advocated [36]. It has been shown that such fixation is unnecessary and offers absolutely no benefit [37].

If bowel is frankly necrotic, resection and a primary anastomosis is performed. In cases where extensive marginal viability of bowel is present, administration of low-molecular-weight dextran (Dextran 40; 10 ml/kg q6h) followed by reexploration in 36 to 48 hours has allowed salvage of ischemic gut that would otherwise have been resected at the original exploration [38]. Prostaglandin E<sub>1</sub> in an experimental small bowel volvulus model appears to have cytoprotective effects during reperfusion of bowel compromised by volvulus [39, 40]. No experience with this or other oxygen-free radical scavenging agents in humans with midgut volvulus has been reported.

Postoperative bowel obstruction is the most frequent complication of a Ladd's procedure, and it is most commonly due to adhesions. Recurrence of volvulus after surgical correction is

rare. Table 3 summarizes the age, sex, the presence of vomiting, volvulus, resection, and mortality of previous reported cases of midgut malrotation.

Malrotation found incidentally in "asymptomatic" children should be corrected to avoid the risk of a supervening volvulus [12]. Biliious vomiting in the pediatric patient, especially the neonate, can be an indication of malrotation with midgut volvulus, and as such it constitutes a true surgical emergency.

### Résumé

La malrotation du tube intestinal est le résultat d'une anomalie embryologique bien connue. Les conséquences d'une malrotation intestinale associée à un volvulus de l'anse intestinale moyenne peuvent être catastrophiques. C'est pourquoi la compréhension parfaite de l'anatomie, des critères diagnostiques et une thérapeutique adaptée sont impératifs. Cet article résume une expérience récente de 18 mois à propos de ce diagnostic, comparé avec les données de la littérature. Plus de la moitié de nos patients (14/22) ont été vus dans le premier mois de vie; tous avaient des vomissements, le plus souvent bilieux. Un transit gastro-intestinal du tube supérieur était l'examen diagnostique préféré, ayant à la fois une sensibilité (18/19, 95%) et une précision 18/21, 86% importantes. Dans cette série, deux-tiers des malades ont présenté un volvulus (15/22, 68%), dont cinq ayant nécessité une résection intestinale en raison d'une ischémie. Un de ces enfants est mort d'un sepsis fulminant. Le procédé de Ladd a été la méthode thérapeutique de choix, qui comprend, selon nous, une éviscération complète, une inspection de la racine du mésentère, une dérotation de l'intestin dans le sens contraire des aiguilles d'une montre, une lyse des bandes de Ladd, redressant le long de la gouttière droite, une appendicectomie avec enfouissement et le placement simple sans pexie du caecum dans la fosse iliaque gauche. Soupçonner cette malformation chez le nouveau-né qui vomit, faire un diagnostic précoce et réaliser une thérapeutique adaptée sont les clés du succès chez les enfants ayant une malrotation intestinale.

## Resumen

La malrotación del tracto gastrointestinal es el resultado de una aberración embriológica bien definida. Puesto que las consecuencias de la malrotación asociada con volvulus del intestino medio pueden ser catastróficas, se hace imperioso poseer un buen conocimiento de la anatomía, los criterios diagnósticos y la adecuada terapia de esta emergencia. El presente informe resume la experiencia reciente en un período de 18 meses con esta entidad y la compara con la publicada en la literatura. Más de la mitad (14/22) de los pacientes se presentaron en el primer mes de su vida y todos exhibieron vómito que, en la mayoría de los casos, era bilioso. La serie gastrointestinal con bario constituyó el estudio diagnóstico de preferencia, y probó ser de alta sensibilidad (18/19, 95%) y certeza (18/21, 86%). En esta serie dos terceras partes de los pacientes se presentaron con volvulus (15/22, 68%), cinco de ellos con necrosis isquémica del intestino que requirió resección. Uno de estos niños murió por sepsis severa. El procedimiento de Ladd fue el tratamiento de elección, el cual, según nuestra propia definición, incluye la evisceración e inspección de la raíz del mesenterio, la desrotación del volvulus del intestino medio en el sentido del reloj, la lisis de las bandas de Ladd con alineación del duodeno sobre la gotera abdominal derecha, apendicetomía y colocación del ciego en el cuadrante inferior izquierdo. Un alto índice de sospecha en el bebé con vómito, la pronta definición del diagnóstico y la instauración de la terapia adecuada resultan en un resultado predeciblemente favorable en los niños con malrotación intestinal.

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