
# Abnormal Embryogenesis As A Pathogenetic Factor Of The Development Of Intestinal Malrotation

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ABSTRACT

Numerous anomalies of bowel and fixation are defined as «incomplete bowel rotation» by many authors. In foreign literature, the term "malrotation" is adopted. This review presents current information on embryogenesis and variants of bowel rotation and fixation anomalies, reveals certain differences in manifestations and teratogenesis depending on its anatomical variant, which can be useful in choosing the tactics of surgical treatment in the neonatal period.

**Keywords:** Embryology, impaired rotation and fixation, malrotation, midgut, SMA.

**Introduction.** Normal rotation of the intestine requires transformation from a simple straight digestive tube to a mature fixed and folded configuration, which is usually formed by the time of birth. Through precise embryological events, the duodeno-jejunal junction is fixed in the left upper abdomen and the cecum is fixed in the right lower quadrant. The midgut, defined as the portion of the intestine supplied by the superior mesenteric artery (SMA), is thus suspended from a broad mesenteric base.

Malrotation is a violation of the normal rotation of any part of the intestinal tract. Literally, malrotation means only “defective” rotation and thus underestimates the importance of improper fixation as a predisposing factor in the development of volvulus. Malrotation is not a single individual entity, but rather a collection of abnormalities that reflect a failure that occurs at any point in midgut development. There are even reports of “hyperrotation”, in which the cecum continues to rotate beyond its normal position, rising upward on the left [11].

The earliest descriptions of intestinal development were made by Mall in 1898 [12] and then expanded upon by Frazer and Robbins in 1915 [5]. Eight years later, Dott translated these preliminary embryological observations into problems that have significant implications in clinical practice [3] in determining the surgical management of malrotation [7]. Ladd described a relatively simple solution to a complex problem [8]. More than 200 postmortem studies had been performed before Ladd's article was published, but he was the first to emphasize the importance of placing the duodenum along the right abdominal wall, thereby widening the base of the mesentery and moving the cecum into the left upper abdomen.

The development of the midgut begins in the fourth week of pregnancy with the differentiation of the colon into anterior, middle and posterior. The mature digestive tract and all associated digestive organs are formed from

this primitive tube. The most common model of midgut maturation involves four distinct stages: 1) embryonal hernia; 2) rotation; 3) retraction and 4) fixation. The intestinal loop can be divided into cranial (duodenojejunal part) and caudal (cecum) ends, which rotate separately but in parallel. In this case, the SMA serves as a fulcrum with the omphalomesenteric duct at the apex. Due to disproportionate growth and elongation of the midgut during the 4th week of pregnancy, the hernia of the intestinal loop turns into an extraembryonic cavity. The intestine then enters a critical period of rotation when the prearterial and postarterial ends make three separate 90° turns, all in a counterclockwise direction around the SMA. The first 90° rotation occurs outside the abdominal cavity. The second 90° rotation begins during the return of the intestines to the abdominal cavity in the 10th week of pregnancy. The duodenojejunal junction now runs posterior to the SMA. The last rotation occurs in the abdominal area. Thus, the primary intestine completes a 270° counterclockwise rotation, allowing the duodenum to be positioned to the left of the SMA, while the caecocolic end is to the right. Then the ascending and descending colon are fixed. Failure to comply with any of these vital steps results in a variety of clinical manifestations of malrotation.

Arrest of embryological development of the midgut can occur at any phase of one or both loops with different consequences [16]. In rare cases, lack of rotation affects only the duodenum and small intestine, while the cecum and colon continue to rotate normally to assume a normal anatomical position [6]. Long et al. [10] reported a case of non-rotation of the colon with normal rotation of the duodenum. “Incomplete rotation” is an arrest occurring during the last 180° counterclockwise rotation of the small bowel and/or the last 180° counterclockwise rotation of the colon. It is this phenomenon that is meant by the term “malrotation” or “mixed rotation”, as well as “partial rotation” [10]. The resulting anomaly ranges from no rotation at all to normal [6]. With abnormal intestinal rotation, the normal attachment of the mesentery is also disrupted, which determines the risk of developing volvulus. In the case of volvulus, the midgut rotates around the axis of the superior mesenteric artery. Twists of 720° or more are often recorded. An increase in the degree of volvulus leads to disruption of intestinal patency, lymphatic, venous drainage and, ultimately, arterial blood supply. Loss of blood supply due to volvulus can lead to catastrophic consequences, including death of the patient. In "reverse rotation", the caudal portion of the midgut first returns to the abdominal cavity, and the duodenum rotates clockwise rather than counterclockwise. As a result, during reverse rotation, the duodenum moves anterior to the SMA rather than posteriorly, and the colon moves posterior to the SMA [6]. Rarely, reverse rotation of the duodenum is accompanied by normal rotation of the colon. This can lead to internal embryonic hernia [6].

Additional abnormalities may result from failure of cecal elongation and failure of small and large intestine fixation. Due to the inability of the cecum to elongate, there is a delay in cecal descent [15]. With incomplete fixation of the ascending colon, a mobile cecum is formed [18]. Because colon elongation occurs and fixation continues during the first months of life, the incidence of a high and/or improperly fixed cecum decreases with age. Impaired fixation of the cecum or sigmoid colon may predispose to the development of cecal or sigmoid volvulus later in life; however, these processes are rare in childhood [18]. Malrotation and improper fixation of the cecum may also predispose the child to intussusception (Waugh syndrome) [1]. A 1985 study by Brereton et al. [1], 41 of 41 children who underwent surgical treatment of intussusception had an “unfixed” cecum. Internal hernias often represent a violation of intestinal fixation [19]. Internal hernia can occur at a variety of other less common sites, also associated with incomplete fusion of the mesentery of the colon [2, 15, 19]. They may be insidious or suddenly manifest as acute obstruction and potential bowel damage [19]. Peritoneal ligaments, commonly known as bands of Ladd, form due to the embryo's disordered attempts to fix the defective intestine. The cords extend from the cecum and

proximal colon to the right subcostal quadrant of the retroperitoneum, often involving the descending and transverse portions of the duodenum. Due to compression by these cords, obstruction of varying degrees, up to complete, can develop. However, most often the bands are present without obstruction or occur only slightly.

Most children with malrotation do not have a predisposing syndrome or genetic predisposition. Malrotation is almost always present in children with congenital diaphragmatic hernia, gastroschisis and omphalocele [4, 9]. The incidence of volvulus in these children is rare, probably due to the anatomy and due to intestinal adhesions that occur after correction of the defect preventing volvulus [9]. Each of these anomalies interferes with the normal spatial development of the intestine. With congenital diaphragmatic hernia, children with a right defect will have greater intestinal deformity than children with a left defect [9].

Historically, the preferred surgical approach for a patient with malrotation has been the Ladd procedure, consisting of laparotomy, release of the midgut volvulus, division of the peritoneal bands obstructing the duodenum, placing the small and large bowel in a nonrotated state, and appendectomy [8, 13]. Today, the survival rate of children with volvulus malrotation is high (>80%), however, despite timely diagnosis and surgical intervention, a significant proportion of patients still die or suffer serious morbidity due to intestinal loss [14, 15, 17].

**Conclusions**. Thus, modern information about embryogenesis and variants of anomalies of intestinal rotation and fixation, certain differences in manifestations and teratogenesis depending on its anatomical variant, may be useful when choosing tactics for surgical treatment of patients in the neonatal period. Paraclinical examinations, especially with vascular imaging, make an important contribution to the correct diagnosis.

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