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"DOLICHOCOLON" EMBRYOLOGICAL BASIS AND ITS CLINICAL IMPORTANCE

Ganesh Elumalai and Manoj P Rajarajan

Department of Embryology, College of Medicine, Texila American University, South America.

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ABSTRACT

A plea is made for the recognition of a disorder of the colon, commonly encountered in the elderly, which is characterized by elongation (and also by dilatation) of the colon, especially the sigmoid. It may give rise to symptoms which suggest a diagnosis of carcinoma. The condition is believed to be acquired, and is an important aetiological factor in the development of sigmoid and, less frequently, of caecal volvulus. Three main colonic transfer forms were identified: slow transit in the proximal colon (STC), normal proximal colonic transit with anorectal retention (NT-AR), and rapid proximal transit ± anorectal retention (RT)

Introduction

The gastrointestinal tract (GIT) rises primarily during the process of gastrulation from the endoderm of the trilaminar embryo (week 3) and extends from the buccopharyngeal membrane to the cloacal membrane. The tract and associated organs later have donations from all the germ cell layers [1-5]. During the 4th week three different regions (fore-, mid- and hind-gut) spread the length of the embryo and will add different mechanisms of the gastrointestinal tract. The large mid-gut is completed by adjacent embryonic folding which "tweaks off" a pocket of the yolk sac, the 2 sections continue to connect through the vitelline duct [4-8]. The oral cavity is formed next breakdown of the buccopharyngeal membrane (oropharyngeal or oral membrane) and donated to mainly by the pharynx lying inside the pharyngeal arches. Loss of buccopharyngeal membrane discharges the tract to amniotic fluid over the remaining of development, and throughout the fetal period is actively accepted. From the oral cavity the next portion of the foregut is originally the pharynx, a single gastrointestinal (esophagus) and respiratory (trachea) common tube, that lies behindhand the heart. Note that the respiratory tract will form from a ventral bud arising at this level. Dolichocolon is the term given to a condition of a long and redundant sigmoid colon. As it occurs in childhood it is associated with obstinate constipation, occasionally a sort of ' overflow diarrhea' and not infrequently acute obstruction from impaction or even volvulus. In general, the prognosis is held to be good, for as the child grows the colon becomes relatively the correct length. No case has been found recorded in which the 'long colon' has developed into the 'mega colon' as described by Hirsch sprung [9-10]. This latter is usually congenital, with constipation and increasing abdominal distension. It is thought to be due to neuromuscular incoordination, especially in relation to the pelvirectal junction. The case here recorded is believed to show characteristics of both these abnormalities of the colon.

Incidence

Dolichocolon was identified in 31 (14.8%) seropositive and 3 (4.8%) seronegative women. The mean length was 57.2 cm in seropositive patients and 52.1 cm in the seronegative patients, that is, the distal colon in Chagas patients was, on average, 5.1cm lengthier. Seropositive female patients existing a mean length of 58.8 cm, and seronegative female patients presented 53.2 cm. Seropositive male patients had a mean length of 55 cm, and seronegative male patients had 49.9 cm [10-12]. Among 191 patients without megacolon and suspected megacolon, the mean length was 56.3 cm in seropositive individuals and 52 cm in seronegative patients. Among individuals with distal colon $\geq$70cm, there were 31 Chagas patients with mean length of 77.9 cm and three seronegative with 71.3 cm. Among 179 with distal colon <70cm, seropositive individuals had a mean length of 53.6 cm, and seronegative patients had 51.2 cm. Serological positive women had longer distal colon than men, whereas the mean length were the same among seronegative individuals [15-16].

Ontogenesis of Dolichocolon:

A. Normal development:

During the first 4-5 weeks of development, the midgut loop forms the physiological umbilical hernia with 90° rotations (first stage) [17-18].

The second stage (10th and 11th week of development) consists of a decrease of the umbilical hernia, with return of the jejunum, ileum, caecum and colon into the abdominal cavity with 180° rotations. As a result, caecum and appendix come to be situated in the sub hepatic region in the right hypochondrium [19-21].

In the third stage (after 10th week of development) descent of caecum and appendix occurs to the right iliac fossa

Tele: +91-9989069546
E-mail address: coolncandy@gmail.com

with the development of the ascending colon and obsession of the mesenteries [21-25].

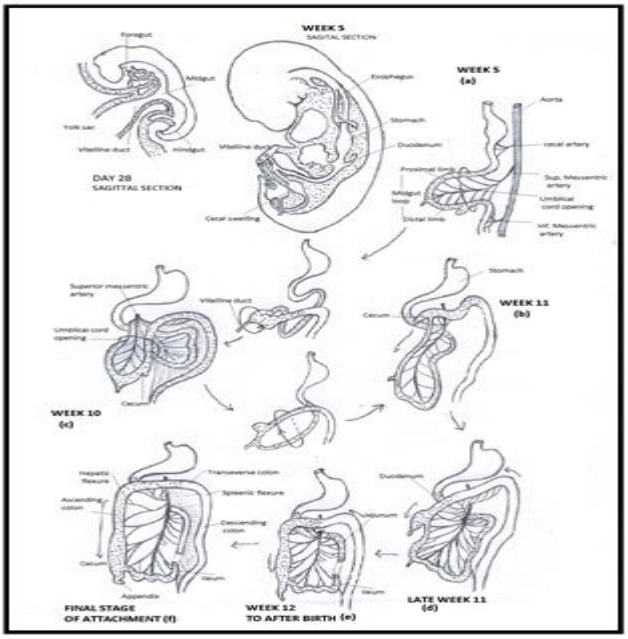

Fig 1. Represents the schematic diagram of normal development of midgut

a)Most of midgut is extruded into base of umbilical cord

(b)All of alimentary track is withdrawn into abdomen.

Cecum lies in epigastrium under stomach

(c)Intestinal elongated hindgut displaced to lift Intra-abdominal intestines comes to lie behind superior mesenteric artery

(d)Colon rotating; Cecum lies in right upper quadrant of abdomen.

e) Rotation of colon is complete; Cecum lies in final position, Common mesentry-mesocolon of ascending colon-continuous with mesentery of ileum

(f) Ascending & descending mesocolon fuse to posterior abdominal wall, jejunum and ileum attach via mesentry to posterior wall from origin of superior mesentry artery to cecum

The midgut loop revolves on the axis of the superior mesenteric vessels through 270° as a whole from its original plane.

Abdominal aorta provides three ventral splanchnic arteries to the digestive tube: the coeliac trunk, the superior arteries and inferior mesenteric arteries. In the fetus as the viscera provided descend into the abdomen, the geneses of these arteries migrate caudally. So the origin of the inferior mesenteric artery is moved from the 12th thoracic vertebra to the 3rd lumbar vertebra and it turns to the left side. This theory can clarify the right-sided course of the inferior mesenteric artery to provide the right sided descending and sigmoid colon in the present case [26-27].

Some fetuses appear to show an extreme elongation of the sigmoid colon and it is possible that perseverance of this condition will give increase to the similar elongation of the colon in the adult life. Such a concept is in accord with the variations happening elsewhere in the gastrointestinal system [28-29].

The hindgut follows the midgut, in the embryo, and spreads from the posterior intestinal portal to the cloacal membrane. It gives growth to the left one-third to one-half or distal part of the transverse colon, the descending colon, the sigmoid colon or pelvic colon, the rectum, the upper portion of the anal canal, and part of the urogenital system. It is provided by the inferior mesenteric artery.

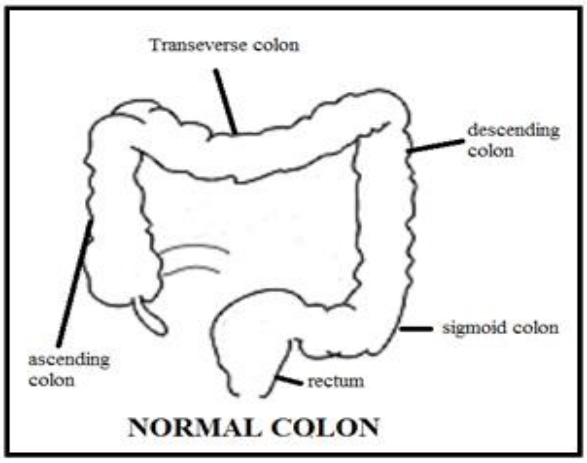

Fig 2. Normal development of colon.

The terminal part of the hindgut passes into the cloaca, which is an endoderm-lined cavity that is in direct communication with the surface ectoderm [30].

B. Abnormal Development

Rotation and fixation initially, the midgut connects with the yolk sac, but this connection narrows to the yolk stalk or vitelline duct. Elongation of the gut occurs faster than elongation of the embryo's body, thus, a series of intestinal variations takes place, usually in 3 stages. Physiologic herniation of the midgut, as it elongates, the midgut forms a ventral U-shaped umbilical loop of gut, the *major intestinal loop*, which schemes into the umbilical cord. This "herniation" takes place at weeks 6 to 10 and is a normal migration of the midgut into the extraembryonic coelom. It occurs because there is not nough room in the abdomen to accommodate the fast-growing midgut due to the space occupied by the massive liver and the kidneys [31-33].

Thus at this stage, the intraembryonic and extraembryonic coeloms connect at the umbilicus, and the midgut develops entirely outside the abdominal cavity.

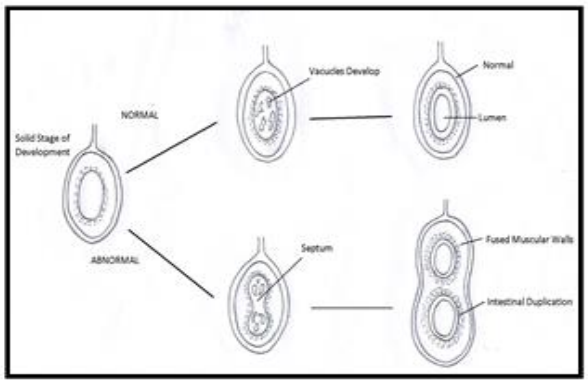

Fig 3. Schematic representation shows the normal and abnormal development of intestine.

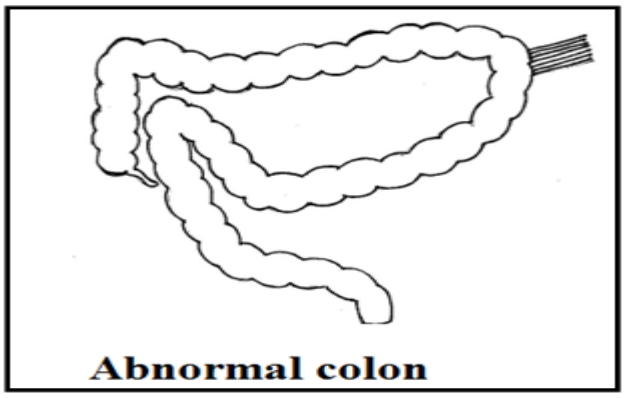

Fig 4. Abnormal development of colon.

The midgut has 2 limbs: a proximal (cranial) and a distal (caudal) lim The yolk stalk is devoted to the apex of the loop at their junction. If the duct persists, it is called Meckel's diverticulum. The proximal limb grows rapidly to form the small intestinal coils (distal duodenum, jejunum, and ileum). The caudal limb changes little except for evolving the lower ileum, the cecal diverticulum, the appendix, the ascending colon and proximal two-thirds of the transverse colon. In the umbilical cord, the midgut loop rotates counter clockwise around the axis of the superior mesenteric artery, taking the proximal limb of the loop to the right and the distal limb to the left. From the artery rise the colic branches for the caudal limb and jejunoileal branches or the proximal limb [33-37]. During the week 10, the intestinal loop will withdraw into the abdomen. The proximal limb (jejunum of the small intestines) revenues first and permits behind the superior mesenteric artery to the left side The later recurring loops settle more to the right. The cecal bulge is seen in the l2 mm embryo as a small conical bulge on the caudal limb of the primitive intestinal loop and is the last part to re-enter the cavity. As they reappear, the gut undergoes another 180^0 counter clockwise rotation, fastening the cecum and appendix near the right lobe of the liver, from where they incline into the right iliac fossa at a later date [38-40]. Return to the abdomen is related to a reduction in relative size of the liver and mesonephric kidneys, as well as abdominal extension and expansion [40-43].

Discussion

Differences in the length and location of any portion of the colon are of developing source and may lead to variety of acute and chronic pathological conditions. One of the most common differences of the sigmoid colon is presence of its terminated loop. Komiyama et al in 1991 informed a 50-year-old Japanese male with extremely long distal part of the colon including the right-sided sigmoid colon. The terminated loop can cause several problems such as constipation, discomfort over the colon, indigestion, loss of weight, insomnia, pain and tenderness in the right iliac fossa.2,4 The symptoms of terminated colon may mimic symptoms of gastric ulcer, heart disease, chronic obstruction of bowel in addition to appendicitis [43-46]. Terminated loop of sigmoid colon is disposed to form a volvulus which can be diagnosed through a CT scan of the abdomen. Chandrika GT have reported a case of right sided sigmoid colon. In the case reported by them, the sigmoid mesocolon prolonged from right iliac fossa to the third sacral vertebra. The current case is unique because of the sigmoid colon lengthening to the left lumbar quadrant of abdomen and layer the left kidney. The connection of the sigmoid mesocolon was also peculiar [47-50]. The N shaped loop shaped by the descending colon and sigmoid colon might lead to confusions in analysis of the barium enema radiographs. This course might cause complications in sigmoidoscopy as well. It might lead to constipation or compressive result on the small intestine or even form a volvulus. Chances of emerging a varicocele are greater with a sigmoid colon abnormality being reported here since it can directly be wrapping the left testicular vessels. The possible cause for this abnormality is the failure of the reordering of the parts of intestine after the reduction of the physiological umbilical hernia during the fetal life [51].

Conclusion

The case here described began as what appeared to be an ordinary dolichocolon: a long, redundant colon with no obvious enlargement of the bowel.

It developed into a typical megacolon of the Hirschsprung type, in which atony and constipation were temporarily relieved by spinal anaesthesia [52]. It throws doubt upon the usually accepted good prognosis of dolichocolon and on the usually accepted etiology of Hirschsprung's disease as a congenital, idiopathic, dilatation of the colon [53]. Diagnosis is established in most cases at laparotomy and is confirmed by histopathology by demonstrating mucosa corresponding to some part of the intestine. Morphologically they are classified into cystic and tubular types. Cystic variety is commoner and usually occurs near ileum. Surgical management includes excision along with a segment of intestine and end to end anastomosis and, marsupialization, opening of the common wall or observation in some asymptomatic cases etc [54]. According to Li et al, excision is possible without resection of the intestinal segment.

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