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"INTESTINAL DUPLICATION" EMBRYOLOGICAL BASIS AND ITS CLINICAL IMPORTANCE

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ABSTRACT

Gastrointestinal duplications are rare congenital anomalies but interesting clinical entities. Most of them showing up in pediatric population they have a varied presentation, with clinical features will vary from asymptomatic abdominal masses to bowel obstruction or perforation. This review traces the embryological origin and describes the anatomical types of duplications. Lack of normal gut motility interferes with movement of intestinal contents In children mostly associated with abdominal surgery or infection. Adhesions are fibrous bands of tissue Cause: postoperative small bowel obstruction after abdominal surgery The majority of obstructions: is single adhesions and can occur any time after 2nd postoperative week.

Introduction

Duplications are uncommon congenital anomalies which can occur anywhere in GIT from mouth to anus. Most of the literature exploration gives only case reports. As Gray AW remarked "gastrointestinal tract is a fertile ground for several and curious congenital malformations which are fascinating to study for both surgeon and pathologist"

There are many theories for growth of duplication. According to one of the theories it is due to persistence of outpouchings of the developing intestines which occur between 6 to 8 weeks of intrauterine life. According to another theory, there is an attachment between the gut endoderm and the neural tube ectoderm, which divides normally in the fourth week of intrauterine development. If this fails, gut will be pulled towards developing vertebral bodies at the site of attachment which results in traction and development of tubular outpouchings, the duplications. If the attachment gets detached at both ends, a duplication cyst results. Theory of aberrant recanalisation is put forward by Bremer in 1944, where he postulates during vacuoles formation and their coalescence to form a single channel, instead of one, two channels form, which may or may not communicate with each other.

Most commonly duplications are classified as two types, the tubular and the cystic varieties. Tubular duplications tend to have communication with the gut lumen commonly at the caudal end but can have communication at both the ends. Cystic type is commoner (65%). Cystic ones are completely separate from the intestine. Li et al have classified small intestinal duplications depending upon the vascular pattern.

Type 1 or Parallel type (74.6%): Duplication is on one of the leaves of the mesentery. Straight artery of the duplication is separate from the straight artery of the bowel. Type 1a: Duplication has a separate mesentery. Type 1b: Duplication shares common mesentery with the bowel. Type 1c: Duplication shares common muscular coat with the bowel. Type 2 or Intra mesenteric type (24.4%): Duplication is located between the two leaves of the mesentery. Straight arteries pass over both the surfaces of the duplication to reach the bowel. Type 2a: Duplication separate from the bowel. Type 2b: Duplication shares common muscular coat with the bowel. They found that vertebral defects were found more commonly associated with type 2 variety (91.6%) than with type 1 variety (5.5%). They claim the study of vascular pattern allows excision of the duplication in small bowel without the need for resection of the small bowel as is commonly practiced. In type 1, mesentery divided from the straight artery and contra lateral artery is kept intact. In type 2, the small branches of the straight arteries are to be divided and the duplication is enucleated. Thus, resection of the bowel avoided resulting in decreased morbidity and operating time

Incidence

It is difficult to estimate the real incidence as many of them may be asymptomatic. It estimated to occur in about 1 in 5000 live births. Over 37 years, 101 duplications are seen in 96 patients in a major children's hospital. From this it appears to be quite rare to see many cases in clinical practice. Seventy-five of these duplications were cystic and 26 were tubular.

Ontogenesis of normal development of intestine

The normal embryology of the midgut is described as it is found in most papers and textbooks of embryology and pediatric surgery. In humans, the development of the midgut starts with the subdivision of the primitive gut into foregut, midgut, and hindgut at the fourth developmental week. At this stage, the midgut is still connected to the yolk sac through the omphaloenteric duct. Many researchers believe that the midgut lies straight in the midline of the embryo in this stage. The Process of Rotation This process can be subdivided into two or three subsequent developmental steps. The early development of the gut anlage into the extraembryonic coelom with a sagittal orientation of the primitive loop (approx. fourth week of development in humans). Many researchers believe that this herniation (physiological umbilical hernia) results because the gut grows too fast in relation to the abdominal cavity of the embryo.

In this stage, the first rotation of the gut anlage inside the extraembryonic coelom takes place. It is 90° in a counterclockwise direction around the axis of the mesentery vessels (approx. eighth week of development in humans). As a result, the midgut loop is now horizontally orientated with the small gut to the right and the colorectum to the left. "Return of the gut" into the abdominal cavity (approx. tenth week of development in humans). At the tenth week of development, the extraembryonic part of the gut enters the abdominal cavity. The details of this process are still unclear. Some authors believe that the process of rotation ends at this stage with another rotation in an anticlockwise fashion (180°). As a result, the flexura duodeni is pushed into a position below and to the left of the root of the mesentery while the cecum and the colon are forced to the right side of the abdominal cavity, thus crossing over the mesenteric root. The end result of these two rotations is a complete rotation of 270°. In the following step, the cecum grows downwards from the upper quadrant of the right abdominal cavity into the right iliac fossa. In contrast to this description, Grob3 subdivides the last rotation of 180° into two steps of 90° each.

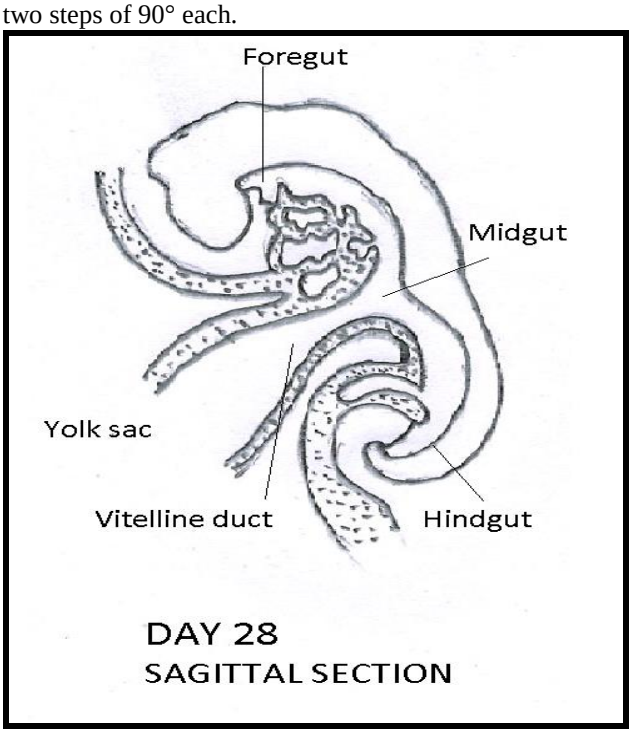

Fig 1. represents the schematic diagram of normal development of midgut.

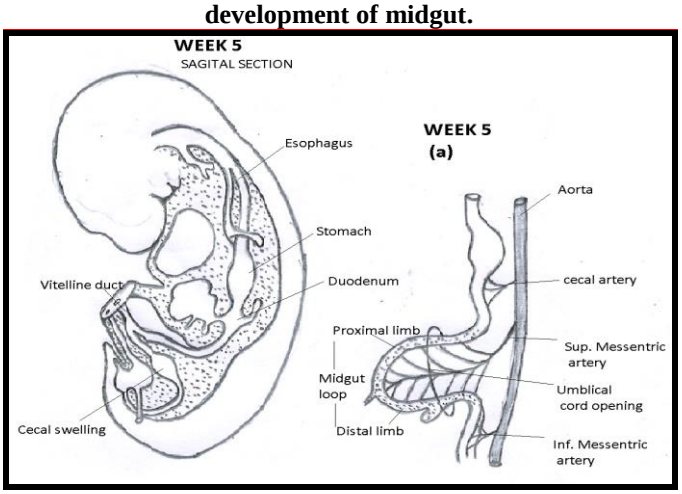

Fig 2. represents the schematic diagram of normal development of midgut in week 5.

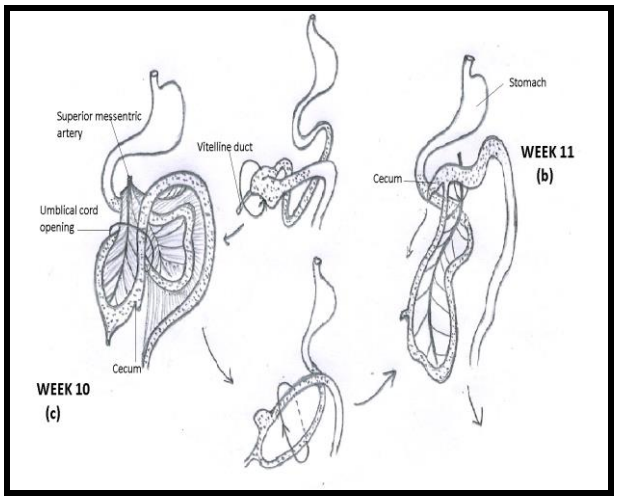

Fig 3. represents the schematic diagram of normal development of midgut in week 10 and week 11

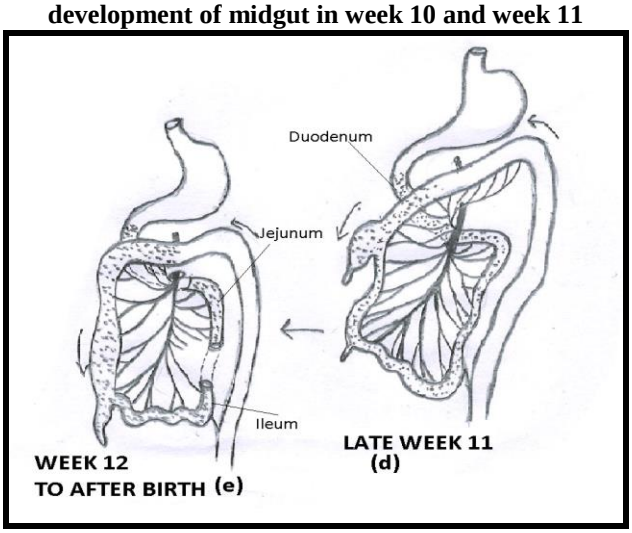

Fig 4. represents the schematic diagram of normal development of midgut in late week 11 and week 12 to after birth.

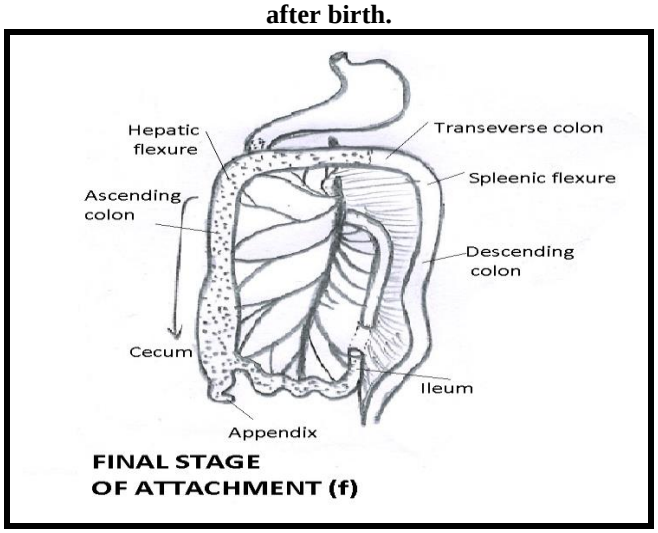

Fig 5. represents the schematic diagram of final stage in normal development of midgut.

Ontogenesis of intestinal duplication

Duplication of the intestines occurs when a segment of the intestines is duplicated as a result of abnormal recanalization (most commonly near the ileocecal valve). The duplication is found on the mesenteric border. Its lumen generally communicates with the normal bowel, shares the same blood supply as the normal bowel, and is lined by normal intestinal epithelium, but heterotopic gastric and pancreatic tissue has been identified.

It is associated clinically with an abdominal mass, bouts of abdominal pain, vomiting, chronic rectal bleeding, intussusception, and perforation.

As with the esophagus and duodenum, the remainder of the intestinal tract is susceptible to various anomalies that seem to be based on incomplete recanalization of the lumen after the stage of temporary blocking of the lumen by epithelium during the first trimester.

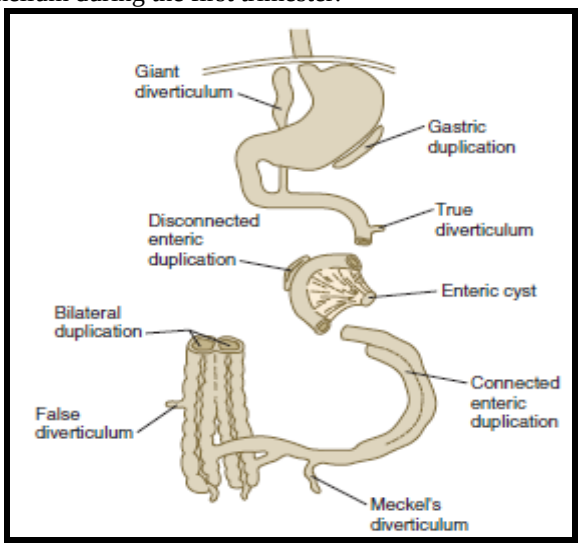

Fig 6. Schematic representation shows the intestinal duplication.

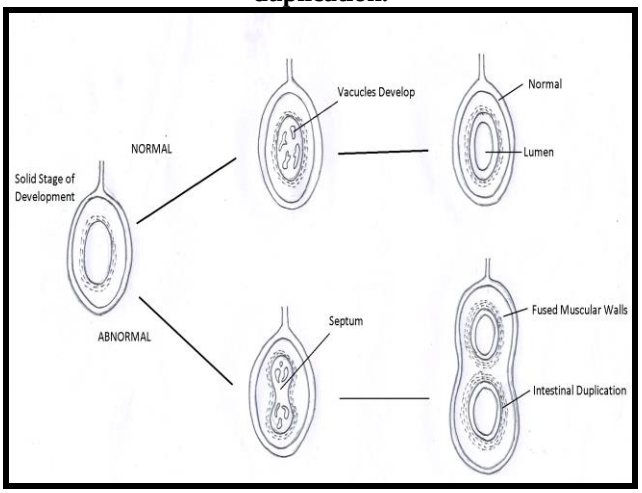

Fig 7. Schematic representation shows the normal and abnormal development of intestine.

Discussion

Duplication of the intestinal tract are rare anomalies consisting of well formed tubular or spherical structures firmly attached to the intestine on the mesenteric side of the lumen. Lined with intestinal mucosa they share a common wall and mesenteric blood supply with the adjacent intestine but usually not communicate with the gut lumen. Potter reported 2 cases in more than 9000 foetal and neonatal autopsies.

The duplication can be classified into localized duplication, duplication associated with spinal cord and vertebral malformation and duplication of the colon. Localized duplications are common in the ileum and jejunum. Theories explain duplication as a defect in recanalisation of the intestinal lumen after the solid stage of embryological development. The split notochord theory proposes neural tube traction mechanism resulting in intestinal duplication along with vertebral and spinal cord anomalies (hemivertebrae, anterior spinabifida, band connection between lesion and cervical or thoracic spine).

The symptoms depend on the size, location and mucosal lining of the cyst. Patient may present with abdominal pain, vomiting, palpable mass or acute gastrointestinal haemorrhage. Intestinal duplication in thorax may present with respiratory distress. Due to nonspecificity, a preoperative diagnosis based on radiography is unlikely. Upper GI study and barium enema demonstrate filling defect or rarely a communication between the cyst and normal bowel.

The ultrasound and CT Scan are useful in establishing diagnosis and may be used to evaluate synchronous lesion once a single duplication has been identified. Duplication of cyst manifest as smooth, rounded, fluid filled cysts or tubular structure with thin slightly enhancing wall on CT Scan. MRI scan shows intracystic fluid with heterogenous signal density on TI weighted image and homogenous high signal intensity on T2 weighted image. Treatment of small cystic or short tubular duplication involves segmental resection along with adjacent intestine. A long tubular duplication cannot be excised as it will lead to short bowel syndrome. In these cases mucosal stripping through a series of multiple incisions is recommended.

According to the review published by Johnson et all in 1994, cancer was found in 3 (23%) of 13 reported cases of ileal duplications in adults (2 adenocarcinoma and 1 squamous cell carcinoma). This evidence of epithelial instability might suggest a tendency toward malignant transformation in long standing duplications. This also supports complete resection of the duplication as the most appropriate method of treatment. Clinical presentation of ATD in adults is variable and, because these lesions occur so infrequently, they are not suspected. A palpable mass can be found in approximately one half of patients; abdominal pain is often present but the most common clinical presentations include intestinal obstruction and bleeding. It is worthwhile to highlight that the clinical presentation is strictly related to the site and type of ATD. In cases of ATD of the hindgut, the diagnosis is often made within the first years of life and the most frequent symptom is biliary vomits. These malformations are usually cystic and localized on the mesenteric border of the first or second duodenum. In the jejunum, the most frequent aspect includes a tubular duplication with a common lumen, whilst in the ileum ADT can resemble a diverticulum. Ileal duplication affecting the distal part of the intestine should be distinguished from a Meckel's diverticulum, even though this is present on the anti-mesenteric border of the intestine. Complications of ATD include volvulus, invagination, bleeding, perforation and malignancy. Twenty-seven cases of ileal duplications in adults are described in the world literature in over 100 years. In one of these cases, the correct diagnosis was made preoperatively. In this case, clinical presentation and pre-operative studies supported a diagnosis of complicated CD. Biopsies were not taken because it was not possible to enter the ileo-cecal valve during diagnostic colonoscopy and the rest of colonic mucosa was normal. Laparotomy is also often indicated in these settings to make a differential diagnosis. Abdominal scans such as SICUS, CT or MRI and conventional contrast x-ray studies are useful tools to detect ATD. The diagnostic problems arise from the extreme rarity of this entity in the adult population. We hereby describe a case of an adult patient who underwent various radiological studies and was referred to different physicians during the year before the correct diagnosis was made. The patient had 3 previous admissions to A&E and was on oral steroids when referred to our

Gastroenterology Unit. ATD was not supposed and the diagnosis was made on the surgical specimen.

Conclusion

Intestinal duplications are rare congenital anomalies presenting clinically most commonly in first 2 years of age. They may be associated with other congenital anomalies like vertebral defects, congenital heart disease, anorectal anomalies etc… Ultrasound and CT scan are helpful in imaging the cyst. Technetium 99 radionuclide scan helps identifying the gastric rests. 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, marsupialisation, opening of the common wall or observation in some asymptomatic cases.

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