 retain 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 omphalenteric 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, Grob 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.

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 T1 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 al 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.

References


[33] Fitz RH. Persistent omphalo-mesenteric remains; their importance in the causation of intestinal duplication, cystoformation, and obstruction. Am J Med Sci 1884; 88: 30-57