CHAPTER 66
Gastrointestinal Duplications

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Introduction

Gastrointestinal duplications (GIDs) are rare congenital malformations. They can arise anywhere from the mouth to the anus.[1] Due to the rarity of these lesions, they frequently present both diagnostic and therapeutic challenges. They may be unexpectedly encountered intraoperatively, and appropriate surgical management requires that the attending surgeon be familiar with the pathology and clinical characteristics of GID.

Demographics

Gastrointestinal duplications occur with a prevalence of 1:4,500 births.[2] In most series, there is no sex predilection. Even though GID is rare, many reports exist with large numbers of patients accumulated over long periods.[3–8] Few reports, however, come from Africa.[9–11]

About 60–70% of patients present before the age of 2 years.[6,8,12] In Africa, due to more difficulties with diagnosis, patients may present at an older age.[11]

Embryology

There are many theories on the embryology of GID. None of these theories, however, is able to explain all types of duplication. The presence of heterotopic tissue in duplications and the consistent mesenteric location of duplications has put many of these theories to question.

These embryological theories include the following:

- *Split notochord syndrome:*[13,14] GID is related to the development of neuroenteric canals, which is related to the thoracic duplications that have associated abnormalities of the cervical and thoracic vertebrae.
- *Abnormalities of recanalisation of the solid stage:*[15] It is only some part of the foregut that undergoes recanalisation, and this process occurs on both the mesenteric and antimesenteric sides of the bowel, whereas duplications are found only on the mesenteric side.
- *Remnants of embryologic diverticula:*[16] This may explain the higher frequency of GID found in the terminal ileum because there are usually numerous diverticula found during development. The presence of heterotopic mucosa, the mesenteric location of GID, and the presence of tubular duplications puts this theory to question.
- *Partial twinning:*[17] This can explain duplications of the hindgut. These are usually associated with malformations of the genitourinary tract.
- *Environmental factors such as trauma or hypoxia:*[18,19] In early foetal development, environmental factors may lead to duplications, and duplications may, in fact, be a part of the spectrum of intestinal atresias.

Pathology

Gastrointestinal duplications are rare congenital malformations. They generally have the following characteristics irrespective of location:

- They are in or adjacent to the wall or part of the gastrointestinal tract and are consistently on the mesenteric side.
- They share a common blood supply with the native bowel.
- They have a definite smooth muscle coat and are lined by alimentary tract mucosa that may be similar to the adjacent bowel or heterotopic tissue.

The most common site of GID is the ileum, followed by the oesophagus, colon, and jejunum (Table 66.1).

Grossly, GIDs are spherical (Figure 66.1) in 82% of the cases; these are the most prevalent type at all levels[20] and do not commonly communicate with the adjacent bowel. The remaining GIDs are tubular, more extensive, more likely to have heterotopic gastric mucosa, and usually communicate with the adjacent bowel, most commonly

Table 66.1: Summary of distribution of locations of 431 gastrointestinal duplications in 395 patients from six studies.

Location	Bower et al. (1978)[6]	Hocking et al. (1981)[7]	Holcomb et al. (1989)[8]	Stringer et al. (1995)[9]	Karnak et al. (2000)[10]	Pulgandla et al. (2003)[11]	Totals (%)
Oropharyngeal	0	–	0	2	1	–	3 (<1)
Oesophagus	15	8	21	15	7	–	66 (15)
Stomach	8	8	8	10	1	6	41 (10)
Duodenum	4	1	2	3	3	7	20 (5)
Jejunum/ileum	34	32	47	21	17	51	202 (47)
Colon	12	4	15	10	9	5	55 (13)
Rectum	2	5	5	6	2	4	24 (6)
Thoracoabdominal	1	2	3	6	2	0	14 (3)
Others	2	0	0	4	0	0	6 (1)
Totals (duplications/ patients)	(78/64)	(60/53)	(101/96)	(77/72)	(42/38)	(73/72)	431 (100)

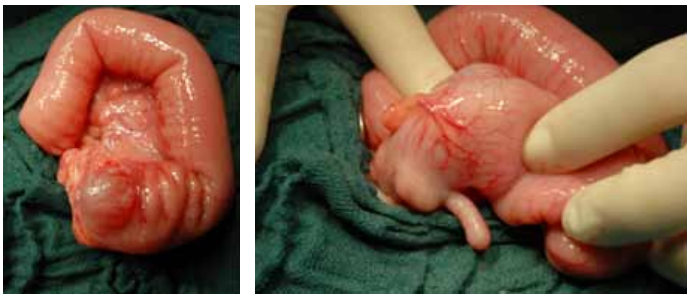

Figure 66.1: Spherical duplication of the ileocaecal area in an 8-month-old child.

in the small and large bowels. The lining mucosa is usually the same as the adjacent normal bowel, but can be heterotopic, such as gastric, squamous, transitional, ciliated columnar mucosa, pancreas, lymphoid aggregates resembling Peyer's patches, and ganglion cells. Others include heterotopic lung tissue or thyroid stroma.[12] However, heterotopic gastric and pancreatic tissues are the ones of significant clinical importance due to the risk of peptic ulceration and pancreatitis. Even though GID in children is benign, malignant transformation has been described in adults.[21]

Diagnosis

Clinical Features

There is no common clinical pattern of signs and symptoms of duplications. They present with a variety of symptoms or sometimes as masses found incidentally during routine examinations or investigations, or they are encountered during an operation for other problems.

Most patients present before the age of 2 years,[11,12] but presentation during adulthood has been described.[4,22] The clinical presentation also varies according to the age of the patient, location of duplication, type of mucosal lining, duration of disease, and presence of complications. The clinical presentation may be due to the pressure effect of the duplication. Feeding difficulties are associated with masses in the floor of the mouth. In thoracic duplications, this leads to respiratory distress or dysphagia. Other symptoms in the chest include recurrent pneumonia and failure to thrive. In the abdomen, GID causes intestinal obstruction but may also cause ureteric, biliary, or even vena caval obstruction. Pancreatitis can arise from pressure on the pancreas. Duplications in the abdomen commonly present with pain, vomiting, and abdominal mass.

The clinical presentation may also be secondary to complications of the duplications. These include intussusception,[12] volvulus,[8] perforation,[23] bleeding (related to ectopic gastric mucosa), peptic oesophageal stricture, and malignant transformation, as seen in adults.[21]

Prenatal Diagnosis

Prenatal diagnosis of GIDs is becoming widespread in the Western world. This ability to accurately identify GID has provided an opportunity to intervene. In cases of nonimmune hydrops in thoracic duplications, thoracoamniotic shunting is carried out in utero in some centres with experience in foetal treatment.[24] This is done in the immediate postnatal period before the onset of symptoms or the development of complications.[2]

Investigations

In the management of GID, accurate preoperative diagnosis is difficult. This is because GID is very rare, and the clinical presentation is nonspecific.

In Africa, where resources are limited, the more expensive investigations, such as computed tomography (CT) scan and magnetic resonance imaging (MRI), should be reserved for the very difficult cases. The most common investigations carried out are ultrasonography (US) and contrast medium examinations.

Ultrasonography

US is the most common modality used and should be the first choice. It typically shows a double-layered wall (inner echogenic mucosa and outer sonoluscent muscular layer). When this double-layered pattern is present on US, a GID is confirmed and there is no need for further radiologic evaluation.[25,26]

Plain X-Rays

A plain chest x-ray (plain abdominal/lateral) will be able to detect foregut duplications in the chest in up to 90% of cases. Plain abdominal x-ray may show evidence of intestinal obstruction.

Contrast Medium Studies

Contrast medium studies may reveal compression or displacement of the adjacent organ. Rarely, it will show communication with the adjacent native organ, but it does not specify the nature of the duplication.

CT Scan or MRI

A CT scan or MRI is employed in difficult cases. It is noninvasive and has the advantage of demonstrating the exact location and relationship to adjacent normal structures. It may also reveal other duplications. It is particularly useful in thoracic, pelvic, and the rare large retroperitoneal duplication cysts. A spinal MRI will outline the relationship of the cyst with the spinal column and spinal canal.

Technetium 99m Pertechnetate Scintigraphy Scan

This scan indicates the definite existence of GID when it contains ectopic gastric mucosa. This is especially useful in oesophageal, duodenal, and tubular small bowel duplications with a high incidence of heterotropic gastric mucosa.

Laparoscopy

Laparoscopy is useful in cases when all the above investigations are not conclusive.

Treatment

The goal is to make a prompt diagnosis and provide treatment before the onset of symptoms or the development of complications. The ideal treatment for GID is complete excision. However, GID in children is a benign disease, and any treatment should not be more radical than to eliminate the patient's complaints and prevent further recurrence.

Important points to be considered in the surgical treatment of GID include:

1. the nature of the blood supply shared between the duplication and native bowel;
2. the presence of heterotopic gastric mucosa, which will negate internal drainage due to the risk of peptic ulceration; and
3. the relationship with adjacent structures, such as the biliary tract in duodenal duplications.

The treatment of GID is best considered by location of the duplication. However, in selected cases, an intraoperative frozen section may give further information on the absence or presence of heterotopic components.

Oropharynx

Oropharyngeal duplications are rare and constitute less than 1% of duplications. They may contain ectopic gastric or colonic mucosa. These cysts are excised by an intraoral incision.

Oesophagus

Oesophageal duplications are related to the right side of the oesophagus and are best approached through a right posterolateral thoracotomy. A supraclavicular approach is used for those located in the cervical region. These lesions should be completely excised due to the high incidence of gastric heterotopia in this location. Where facilities are available, a thoracoscopic approach is preferable for isolated lesions.[27]

Thoracoabdominal

Thoracoabdominal cysts are not intimately adherent to the oesophagus but are usually to the posterior right side of the chest and may have communication through the right crus of the diaphragm to the pylorus, duodenum, jejunum, or ileum. They are often lined by ectopic gastric mucosa. It is also important to ensure that there is no neuroenteric communication. If present, this should be excised in consultation with a neurosurgeon. Incomplete excision will lead to meningitis. The best approach here is the use of separate posterolateral thoracotomy and abdominal approaches. Depending on the extent of the lesion and the patient's condition, it may be carried out at the same surgery or staged. For isolated lesions, thoracoscopic resection is becoming the preferred treatment.

Gastric

Gastric duplications represent 8% of duplications. Unlike other duplications, these are more common in girls.[28] Most of these duplications are on the greater curvature, but they can rarely be on the lesser curvature or the pylorus. They usually do not communicate with the stomach, and complete excision is possible. More extensive lesions are excised with a limited partial gastrectomy.

Duodenum

Duodenal duplications also are rare. The treatment of choice for duodenal duplications is complete excision with preservation of the duodenum. However, this total excision is not always possible if the cyst is in close proximity to the pancreas or the biliary or pancreatic ducts.

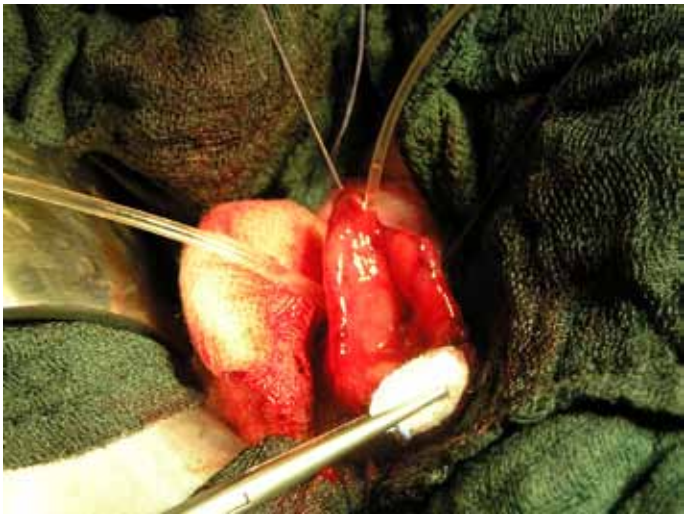

Figure 66.2: Communicating cystic duplication of the duodenum. Exposure after duodenotomy. The upper tube is within the major papilla; the lower one intubates the cyst.

The walls of the ducts may be included in the thickness of the wall of the duplication and may also share the same blood supply with the cyst (Figure 66.2). In these cases, the options include:[29]

- partial resection of part of the cyst wall but including the mucosal lining; or
- internal drainage through a wide cystoduodenal anastomosis or a cystojejunal Roux-en-Y anastomosis.

Small Bowel

The small bowel (jejunum/ileum) is the site of about half of all GIDs. The cystic types of duplications are sometimes easily shelled out, but most are resected with primary end-to-end anastomosis to restore bowel continuity. A laparoscopic approach may be effective to identify the lesion and minimize the abdominal wall incision by lifting up the affected bowel segment.

The tubular type of GID can involve the whole ileum. It has an 80% incidence of gastric heterotopia. Extensive resections will lead to short bowel syndrome, and drainage into the adjacent normal bowel is not encouraged due to the risk of peptic ulceration. In these cases, the multiple stepwise stripping of the mucosa is as described by Wrenn,[30] but anastomosis of the duplication to the stomach has also been tried.[31]

Colon

Colonic duplications rarely contain ectopic gastric mucosa. The rare complete colorectal duplication may be associated with doubling anomalies of the genitourinary organs, such as the bladder and vagina.[17] Cystic duplications can be shelled out or resected with primary colocolostomy. Tubular duplications of the colon have one or more communications with the native bowel. If it is extensive, a distal communication is created with the colon or rectum.

Rectum

Rectal duplications are usually presacral. In 90% of cases, there is no communication with the rectum. The posterior sagittal approach is preferred. It gives good exposure, facilitates safer removal and repair, and prevents entering or compromising the rectal lumen. Laparotomy is preferable, however, for the rare anterior duplications.[32]

Prognosis and Outcome

The outcome of surgical treatment of duplications is good. Poor outcomes are observed when there are associated severe malformations, which in themselves carry a high morbidity and mortality.

Evidence-Based Research

GIDs are very rare anomalies, so most large series are accumulated over three to four decades from big referral centres. It is difficult to have randomized studies.

Key Summary Points

1. Gastrointestinal duplications are rare.
2. GIDs can arise from the mouth to the anus.
3. The aetiology of gastrointestinal duplications is heterogenous.
4. About half of all duplications are located in the small bowel.
5. The clinical features of gastrointestinal duplications is nonspecific; a high index of suspicion is required for prompt diagnosis.
6. Most patients with gastrointestinal duplications will present before 2 years of age, but presentation during adulthood, as well as late malignant transformation, has been described.
7. The presence of ectopic gastric mucosa, especially in tubular duplications, should be documented, as it has an impact on the approach to management.
8. Ultrasonography should be the first line of investigation.
9. The ideal treatment is total excision; however, in children, GID is a benign disease, and therapy should not jeopardise the integrity of the adjacent normal bowel.
10. The results of surgical treatment are good.

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