Spectrum of complications related to congenital and developmental abnormalities of the gastrointestinal tract in adolescents and adults: Evaluation with cross sectional imaging

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Learning objectives

1. Understand the embryology of the gastrointestinal tract.

2. Discuss the various complications associated with congenital and developmental abnormalities of the gastrointestinal tract in the adolescents and adults.

3. Recognize imaging features of various complications related to congenital and developmental abnormalities of the gastrointestinal tract in the adolescents and adults.

Background

A wide spectrum of congenital anomalies can affect the gastrointestinal (GI) tract. Most symptomatic congenital anomalies in the GI tract are found in newborns and young infants. However, some may present in adults, which can be explained by some patients with congenital anomalies of the GI tract remaining asymptomatic and the condition being discovered incidentally by imaging studies or at surgical exploration or even autopsy. Other patients may have milder forms of disease that are diagnosed in newborns and young infants and then go unnoticed until later in their lives, such as congenital esophageal stricture, adult Hirschsprung disease, annular pancreas, or intraluminal duodenal diverticulum. Others may have delayed symptomatic manifestations, due to secondary changes associated with congenital anomalies in the GI tract, such as volvuli related to abnormal fixation of the hollow viscus and neoplastic or inflammatory changes of the heterotopic pancreas.

Radiologic examinations play a useful role in the evaluation of patients with congenital anomalies of the GI tract. Various anomalies of the GI tract are increasingly being discovered by computed tomography (CT), because of the more widespread use of CT for the evaluation of adult patients suspected of having non-specific abdominal symptoms or acute abdomen. Thus, identification of a variety of symptomatic congenital anomalies in the GI tract by CT may be important in the determination of appropriate treatment.
In this article, we review the embryologic development of the GI tract. We also discuss and illustrate various complications associated with congenital and developmental abnormalities of the GI tract, including congenital esophageal stricture, gastric volvulus, duodenal web, annular pancreas, heterotopic pancreas, midgut volvulus, cecal volvulus, anomalies of the omphalomesenteric duct, Hirschsprung disease, and duplication. Additionally, we briefly comment on internal hernias.

**Imaging findings OR Procedure details**

**Embryogenesis of the gastrointestinal tract**

*Development of midgut*

During the 6\textsuperscript{th} to 9\textsuperscript{th} weeks of gestation, the abdominal cavity cannot accommodate the rapidly expanding liver and GI tract. As a result, the primitive midgut herniates into the area of the yolk stalk ("physiological umbilical herniation") with a 90\textdegree counterclockwise rotation of the intestine around an axis formed by the superior mesenteric artery (SMA). During the 10\textsuperscript{th} to 12\textsuperscript{th} weeks, the primitive midgut, with an additional 180\textdegree counterclockwise turn, has completely re-entered the abdominal cavity. The cecum is initially located just inferior to the liver. By about the 12\textsuperscript{th} week of gestation, the cecum starts to descend into the right iliac fossa, placing the ascending colon on the right side of the abdominal cavity. The ascending colon, except for its most caudal part, fuses the retroperitoneum. Finally, the duodenum forms a C-loop and is fixed to the retroperitoneum in the left upper quadrant of the abdomen at the ligament of Treitz, and the cecum becomes similarly fixed to the retroperitoneum in the right lower quadrant. Normal small bowel mesentery has a broad attachment, extending from the duodenojejunal junction in the left upper quadrant to the cecum in the right lower quadrant (*Fig 1*) (1).

The omphalomesenteric or vitelline duct connects the primitive midgut with the yolk sac. During the 5\textsuperscript{th} to 7\textsuperscript{th} weeks of gestation, the omphalomesenteric duct normally regresses, as the placenta replaces the yolk sac as the main source of fetal nutrition (*Fig 1*) (2).
Development of the pancreas

The pancreas develops from a single dorsal and two ventral buds, originating from the endodermal lining of the duodenum during the 4th to 5th weeks of gestation. Two ventral buds are derived from the hepatic diverticulum, whereas one dorsal bud arises from the dorsal mesogastrium. The left ventral bud regresses before rotating, and then the right ventral bud rotates dorsally to the left of the duodenum and fuses with the dorsal bud by about the 7th week of gestation. The ventral bud becomes the uncinate process and pancreatic head, and the dorsal bud forms the body and tail. The main pancreatic duct (of Wirsung) is formed by the distal part of the dorsal pancreatic duct and the entire ventral pancreatic duct. The proximal part of the dorsal pancreatic duct may persist as the accessory pancreatic duct (of Santorini) (3).
Congenital esophageal stenosis

Congenital esophageal stenosis is a rare developmental anomaly. Symptomatic congenital esophageal stenosis is usually present in newborns and young infants. Much less frequently, less severe forms of stenosis may present later in adults. Most present with dysphagia, vomiting, or respiratory symptoms, although symptoms depend on the location and the severity of the stenosis. Congenital esophageal stenosis is characterized by three forms, stenosis due to tracheobronchial remnants, fibromuscular stenosis, or a membranous diaphragm. The etiology of congenital stenosis is uncertain. Stenosis due to tracheobronchial remnants may be caused by defective embryologic separation of the primitive foregut from the respiratory tract, and can be associated with esophageal atresia and tracheoesophageal fistula. Esophageal stenosis due to tracheobronchial remnants usually involves the lower third, whereas fibromuscular stenosis and membranous diaphragm are common in the middle third. However, esophageal stenosis can affect any portion of the esophagus (4).

On an esophagogram, congenital esophageal stenosis is characterized by a segmental stricture with a smooth tapered area of concentric narrowing in the esophagus, and is often associated with ring-like constriction, which may be due to tracheobronchial remnants. CT may also demonstrate segmental stricture with circumferential wall thickening and smooth tapering (Fig 2) (4).
Fig.: Figure 2. Congenital esophageal stenosis (fibromuscular form) in a 25-year-old man with a 4-year history of dysphagia. (a) Esophagogram shows focal concentric narrowing (arrow) of mid thoracic esophagus with proximal dilatation.

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Fig.: Figure 2. Congenital esophageal stenosis (fibromuscular form) in a 25-year- old man with a 4#year history of dysphagia. (b) Coronal reformatted CE-CT shows circumferential wall thickening in the region of the esophageal stricture (arrows). Proliferation of smooth muscle with chronic inflammation was confirmed at myotomy. 

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

Esophageal strictures caused by trauma, reflex esophagitis, or ingestion of toxic substances may have a similar appearance. The clinical history can be helpful for differentiating the various causes of esophageal stenosis, because exclusion of other diseases known to be associated with esophageal stricture in adults is important to suggest congenital esophageal stenosis (4).

Gastric volvulus
Gastric volvulus, defined as an abnormal torsion of the stomach, usually more than 180°, is a rare condition that is encountered in the adult as well as the pediatric age group. It presents either as an acute abdominal emergency or with chronic, intermittent symptoms, depending on the anatomical orientation, the rapidity of onset, the degree of rotation, and the degree of obstruction. Chronic intermittent symptoms are more common than the acute form. Acute and complete gastric volvulus is a surgical emergency, because of the increased risk of ischemia with secondary necrosis, perforation, and shock (5, 6).

Gastric volvulus is usually classified into two main types, mesenteroaxial and organoaxial (Fig 3). Mesenteroaxial volvulus is much less common than organoaxial volvulus, and is most often seen in children. The mesenteroaxial type occurs when the stomach rotates on its short or perpendicular axis. It is thought to be the result of abnormal laxity or the absence of peritoneal attachments of the stomach, due to abnormal fusion of the fetal mesenteries, and can also be associated with wandering spleen. In comparison, organoaxial volvulus is more common and is usually seen in adults. It is often seen in conjunction with a diaphragmatic defect, such as hiatal hernia, diaphragmatic eventration, and diaphragmatic rupture. This defect results in abnormal migration of the stomach into the thorax and can allow the stomach to rotate on its long axis, connecting the cardia with the pylorus (5, 6).
Fig.: Figure 3. Gastric volvulus. (a) Organoaxial gastric volvulus.

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Fig.: Figure 3. Gastric volvulus. (b) Mesenteroaxial gastric volvulus.

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

Although a fluoroscopic study is the gold standard for the diagnosis of gastric volvulus, multidetector CT with three-dimensional volume rendering and multiplanar reformation is often performed in the setting of acute abdomen and can be helpful in the evaluation of gastric rotation, characterization of volvulus type, and detection of other associated abnormalities, such as gastric ischemia, hiatal hernia, or wandering spleen. CT findings of gastric volvulus may be variable, depending on the extent of diaphragmatic herniation, the point of torsion, and the final position of the stomach. In organoaxial volvulus, CT demonstrates an "upside-down" stomach, with the lesser curvature below the greater curvature. A low gastroesophageal junction and distortion of the duodenum are also seen on CT. In mesenteroaxial volvulus, CT demonstrates that the pylorus has rotated from right to left, so that the stomach lies "right-side up" (Fig 4) (5, 6).
**Fig.** Figure 4. Perforated gastric volvulus (mesenteroaxial type) with emphysematous gastritis in a 25-year-old woman with abdominal pain. (a) CT scanogram shows gastric distention and pneumoperitoneum (*).

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
**Fig.**: Figure 4. Perforated gastric volvulus (mesenteroaxial type) with emphysematous gastritis in a 25-year-old woman with abdominal pain. (b, c) Axial contrast-enhanced CT images show a distended stomach with an anterior location of the gastric antrum (arrow in b) related to the gastric fundus. Note collections of intramural gas within the stomach wall (arrowheads) and pneumoperitoneum (*). The spleen with a partial infarct is displaced medially. F=gastric fundus, B=gastric body, S=spleen.

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**Fig.** Figure 4. Perforated gastric volvulus (mesenteroaxial type) with emphysematous gastritis in a 25-year-old woman with abdominal pain. (b, c) Axial contrast-enhanced CT images show a distended stomach with an anterior location of the gastric antrum (arrow in b) related to the gastric fundus. Note collections of intramural gas within the stomach wall (arrowheads) and pneumoperitoneum (*). B= gastric body, A=gastric antrum.

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
**Fig.:** Figure 4. Perforated gastric volvulus (mesenteroaxial type) with emphysematous gastritis in a 25-year-old woman with abdominal pain. (d) Coronal reformatted contrast-enhanced CT (lung window setting) shows twisted stomach with intramural gas (arrowheads). Note pneumoperitoneum (*). F=gastric fundus, B=gastric body, A=gastric antrum.

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

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**Duodenal web**

Congenital duodenal anomalies such as atresias or webs are rare developmental anomalies caused by a failure of the recanalization process in the primitive foregut, between the 8\textsuperscript{th} and 10\textsuperscript{th} weeks of gestation. Congenital duodenal anomalies have often
been associated with additional congenital anomalies, such as annular pancreas, midgut malrotation, imperforate anus, and Down's syndrome (7).

There are several types of webs, including complete duodenal atresias or imperforate webs, imperforate intraluminal duodenal diverticulum (IDD) or windsock webs, and perforated duodenal webs or IDD, with either central or eccentric opening. IDD occurs as a saccular structure, which is connected to either the entire circumference or only part of the wall of the duodenum (eccentric opening) at or near the ampulla of Vater. Perforated duodenal webs or IDD with central or eccentric openings may present in adults with symptoms associated with pancreatitis, peptic ulcer, and duodenal obstruction. The delayed onset of duodenal obstructive symptom in adults with IDD is difficult to explain and seems to be an effect of the progressive loss of compensatory peristaltic action to overcome the small duodenal aperture. The pathogenesis of pancreatitis in patients with IDD is also unclear. However, reflux of the duodenal contents through the papilla of Vater is the most widely accepted proposed etiology (7).

The diagnosis of the IDD can be made endoscopically, although the endoscopic view can be misinterpreted as scarring from peptic ulcer lesions. On fluoroscopic examination, IDD typically appears as a barium-filled "windsock" deformity, originating from the second portion of the duodenum. The shape of the wall of the IDD can be changed by peristalsis in the same way as a windsock. The diagnosis can be made with CT or MR, where fluid- or contrast material-filled IDD and thin diverticular walls may be demonstrated (Figs 5-7) (7).
**Fig.**: Figure 5. Intraluminal duodenal diverticulum in a 22-year-old woman with heterotaxy syndrome (polysplenia) who presented with abdominal pain. (a) Axial contrast-enhanced CT shows a midline liver, multiple spleens (arrows) in the right upper quadrant adjacent to the stomach, and IVC interruption with azygous continuation (arrowhead).

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Fig.: Figure 5. Intraluminal duodenal diverticulum in a 22-year-old woman with heterotaxy syndrome (polysplenia) who presented with abdominal pain. (b) Axial contrast-enhanced CT at slightly more caudal than (a) shows a thin rim (arrowheads), corresponding to the wall of the intraluminal duodenal diverticulum.

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Fig.: Figure 5. Intraluminal duodenal diverticulum in a 22-year-old woman with heterotaxy syndrome (polysplenia) who presented with abdominal pain. (c) Oblique sagittal reformatted CT image shows blind-ending saccular structure with the "windsock" appearance (arrow) within the second portions of the duodenum.

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
**Fig.**: Figure 5. Intraluminal duodenal diverticulum in a 22-year-old woman with heterotaxy syndrome (polysplenia) who presented with abdominal pain. (d, e) Images from virtual endoscopy (d) and conventional endoscopy (e) show an eccentric opening (arrow) in perforated duodenal web and food impaction (*) within a duodenal diverticulum.

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Fig.: Figure 6. Intraluminal duodenal diverticulum in a 31-year-old woman with acute pancreatitis. (a) Upper gastrointestinal series image shows a contrast-filled intraluminal duodenal diverticulum with the "windsock" appearance (arrowhead). Note the radiolucent wall that separates the diverticular lumen from the remainder of the duodenal lumen. (case courtesy of J.Y. Kim, M.D., Kyungpook National University Hospital, Daegu, Korea)

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Fig.: Figure 6. Intraluminal duodenal diverticulum in a 31-year-old woman with acute pancreatitis. (b) Oblique coronal reformatted image shows a debris-filled "windsock" intraluminal diverticulum (arrowheads) distorting the pancreas and the second and third portions of the duodenum. Note mild dilatation of the main pancreatic duct (arrow). (case courtesy of J.Y. Kim, M.D., Kyungpook National University Hospital, Daegu, Korea)

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
**Fig.**: Figure 7. Intraluminal duodenal diverticulum with situs anomaly in a 32-year-old man with hematochezia. (a) Coronal reformatted contrast-enhanced CT image shows two intramural diverticula with "windsock" appearance (arrows) in the 3rd and 4th portion of the duodenum.

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Fig.: Figure 7. Intraluminal duodenal diverticulum with situs anomaly in a 32-year-old man with hematochezia. (b) Upper gastrointestinal series image shows two contrast-filled intraluminal duodenal diverticula with the "windsock" appearance (arrows).

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Annular Pancreas

Annular pancreas is a rare congenital anomaly in which pancreatic tissue encircles the second portion of the duodenum. Two major theories have been proposed concerning the development of annular pancreas. Lecco suggested that the right ventral bud adheres to the duodenal wall and becomes stretched and elongated following rotation. According to Baldwin, the left ventral bud persists and migrates around the duodenum in opposite directions, to fuse with the dorsal pancreatic bud, resulting in an annular pancreas (Fig 8). Both theories have their limitations and cannot explain all anatomic types of ductal arrangement in annular pancreas (3, 7). The presentation in adults differs from that in children. Annular pancreas usually presents during the first year of life, with symptoms...
due to gastric outlet obstruction; this condition can manifest in adults with peptic ulcers, duodenal obstruction, pancreatitis, or periampullary carcinoma. However, approximately 50% of cases may be asymptomatic for life, with the anomaly detected incidentally (3).

**Fig.:** Figure 8. Formation of the annular pancreas

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

Imaging modalities are important in establishing the diagnosis of annular pancreas, although surgery remains the diagnostic gold standard. The diagnosis of annular pancreas can be established by ERCP, which allows for exact delineation of the aberrant pancreatic duct encircling the duodenum. More recently, annular pancreas has been diagnosed with CT, MR, and MRCP. CT and MR imaging show the pancreatic parenchyma, completely or partially encircling the descending duodenum at or below the ampulla of Vater (**Fig 9**).
**Fig.**: Figure 9. Annular pancreas in a 59-year-old man with repeated episodes of vomiting. (a) Coronal thick-slab single-shot MRCP shows aberrant pancreatic duct (arrow) encircling the descending portion of the duodenum with dilatation of the proximal duodenum (*). Note mild dilatation of main pancreatic duct (arrowhead).

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Fig.: Figure 9. Annular pancreas in a 59-year-old man with repeated episodes of vomiting. (b) Axial gadolinium-enhanced T1-weighted three-dimensional gradient echo image obtained during the late arterial phase shows pancreatic tissue (arrows) incompletely encircling the descending portion of the duodenum.

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Fig.: Figure 9. Annular pancreas in a 59-year-old man with repeated episodes of vomiting. (c) Endoscopic image shows stenosis of the descending duodenum (arrow).

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

Secretin-enhanced MRCP allows better visualization of the aberrant pancreatic duct encircling the duodenum. An annular pancreatic duct may drain into the intrapancreatic common bile duct, the duct of Wirsung, or the duct of Santorini. Pancreas divisum is often found in the presence of annular pancreas. CT and MR imaging are also useful in delineating coexistent abnormalities, such as pancreatitis and periampullary carcinoma (3, 7).

**Heterotopic pancreas**

Heterotopic pancreas arises with the detachment of one or more branching buds during embryonic rotation and fusion of the dorsal and ventral pancreatic buds. Heterotopic pancreas occurs in 1-14% of the population. Heterotopic pancreas is usually
asymptomatic, although complications can develop, including pancreatitis, pseudocyst, cystic dystrophy, insulinoma, and malignant transformation (3).

Heterotopic pancreas is usually located in the submucosa and/or muscularis propria, particularly along the greater curvature of the gastric antrum or in the proximal duodenum. On contrast-enhanced CT, it usually appears as a homogeneously enhancing intramural subepithelial mass, less than 3 cm in diameter, with an irregular border, flat or ovoid shape, and intraluminal growth. The degree of enhancement is usually similar to that of the pancreas (3).

**Cystic dystrophy** is an uncommon complication of heterotopic pancreas. It most often develops in the wall of the second part of the duodenum and is characterized by the presence of cystic formation, surrounded by inflammation and fibrosis in the heterotopic pancreas. In contrast to pseudocyst, the inner surface of cystic dystrophy is lined with columnar epithelium, similar to that lining the pancreatic duct. The pathophysiology of cystic dystrophy of heterotopic pancreas remains unclear. Alcohol and mechanical obstruction of heterotopic pancreatic ducts have been proposed in its pathogenesis. Contrast-enhanced CT shows two or more, relatively small sized (often 1-1.5 cm in diameter), and round-shaped cystic lesions within the markedly thickened duodenal wall ([Fig 10](#)). Strong enhancement of lesions in the duodenal wall is often demonstrated, because of associated inflammatory and fibrotic changes. However, the imaging features of cystic dystrophy may be similar to those of pseudocyst that develop in the duodenal wall or the pancreaticoduodenal groove area (8).
**Fig.**: Figure 10. Cystic dystrophy of heterotopic pancreas in a 56-year-old man. Coronal reformatted CT cholangiogram shows multiple cysts (arrows) along the thickened duodenal wall. The duodenal lumen is displaced and compressed laterally. (case courtesy of J.Y. Oh, M.D., Dong-A University Hospital, Busan, Korea)

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**Midgut malrotation with volvulus**

Midgut malrotation includes a spectrum of congenital abnormalities of intestinal position and fixation, and refers to either nonrotation or incomplete rotation of the midgut around the axis of the SMA during fetal development. Reversed rotation can occur, but is extremely rare. It is usually an isolated abnormality, but it can be associated with other congenital anomalies, such as inferior vena cava anomalies, polysplenia, short pancreas, and preduodenal portal vein (1).
Symptoms in patients with malrotation are caused by volvulus, obstruction from peritoneal bands, rarely internal hernia, or a combination of both. Midgut malrotation with volvulus typically presents during the first year of life, with symptoms of bilious vomiting and intestinal obstruction, but may also present in adulthood. In adults, the clinical presentation of malrotation is usually insidious and can often be mistaken for other common abdominal diseases, because this condition is relatively rare in adults and also often manifests as chronic abdominal complaints or nonspecific symptoms, secondary to intermittent volvulus. However, acute episodes may occur without a history of intestine-related problems in a few adult patients (1, 6).

Malrotation is always accompanied by abnormal fixation, in the form of a peritoneal (Ladd's) band. Ladd's bands arise in the right upper quadrant, due to a malpositioned cecum in the upper quadrant of the abdomen, and across the second and third portions of the duodenum to the right paracolic gutter. Duodenal obstruction may result from compression by peritoneal fibrous bands (1).

Malrotation is usually associated with abnormal fixation of the small bowel mesentery, which results in an abnormally short small bowel mesenteric root. This allows abnormal mobility and torsion of the midgut, which can ultimately lead to midgut volvulus. Midgut volvulus causes intestinal obstruction and possibly ischemia of the bowel, due to superior mesenteric vascular compromise. Thus, midgut volvulus can be a life-threatening condition requiring immediate surgical intervention (1, 6).

The upper GI series is usually the preferred diagnostic test for midgut malrotation with or without volvulus, particularly in infants. Fluoroscopic images of malrotation may show a vertical duodenum that does not cross the midline, with most of the small bowel in the right abdomen. Upper GI series in midgut volvulus show a typical corkscrew-like appearance of the twisted segment of jejunoileal loops and duodenal or small bowel obstruction, as well as the malrotated bowel (1, 6).

As the use of CT in clinical practice has increased in recent years, malrotation with or without volvulus has increasingly been detected in adults. CT features of malrotation include an inability to see the third part of the duodenum across the midline, from right to left, a right-sided small bowel, a left-sided colon, and the reversal of the normal relationship between the SMA and the superior mesenteric vein (SMV) (Fig 11) (1, 6). If the SMV lies to the left of or posterior to the SMA on cross-sectional imaging, malrotation should be considered. However, reversal of the normal relationship of superior mesenteric vessels can be seen in patients without malrotation, and a normal relationship of superior mesenteric vessels can be found in patients with malrotation.
Thus, an inability to see the third part of the duodenum cross the midline, from the right to left side, and the identification of abnormal bowel position is more important. The whirlpool sign or whirl-like pattern on CT is the result of wrapping of the SMV, tributaries of the SMV, mesenteric fat, bowel, and branches of SMA around the SMA, in a clockwise direction, and can be highly diagnostic of midgut volvulus (Fig 11) (6). This distinctive whirl pattern can also be demonstrated on ultrasound. However, the whirl sign may be associated with internal hernia or adhesion, and can also be seen after prior surgery, due to disruption of the normal anatomic relationships of vessels and bowel loops; thus, the whirl sign is not a specific finding for midgut volvulus (Fig 12).

**Fig.:** Figure 11. Midgut volvulus in a 68-year-old man with acute abdominal pain. (a) Contrast-enhanced CT shows superior mesenteric vein (arrow) lying to the left to the superior mesenteric artery (arrowhead) (reversal of the normal relationship between superior mesenteric artery and superior mesenteric vein).
Fig.: Figure 11. Midgut volvulus in a 68-year-old man with acute abdominal pain. (b) Contrast-enhanced CT at slightly more caudal than (a) shows the whirl pattern (arrows) of jejunal loops encircling the superior mesenteric artery (arrowhead).

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
**Fig.:** Figure 12. Whirl sign associated with postoperative adhesion in a 55-year-old man who underwent small bowel resection due to traumatic injury 22 years earlier. Axial contrast-enhanced CT shows the "whirl appearance" (arrows) around the superior mesenteric artery. Note normal position of the ascending and descending colon.

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

Additionally, CT is useful in depicting the complications of volvulus, such as the degree of small bowel obstruction and the presence of ischemia or edema in the bowel, and other associated congenital anomalies with midgut malrotation (1).

**Cecal and sigmoid volvulus**
**Cecal volvulus** accounts for approximately 40-50% of all cases of colonic volvulus. Cecal volvulus usually occurs in patients who have excess mobility of the right colon as a result of congenital incomplete fusion of the mesentery of the right colon with the retroperitoneum. Acquired conditions, such as a pelvic mass, adhesion from prior surgery, pregnancy, colonic atony, and recent colonoscopy can also contribute. It usually occurs in much younger patients than those seen for sigmoid volvulus, i.e., 30-60 years. Most patients usually present with a gradual onset of acute cramping pain, distension, constipation, and vomiting, similar to the symptoms of small bowel obstruction. Bowel ischemia and perforation may occur as complications. Thus, cecal volvulus is a surgical emergency (6, 9).

Cecal volvulus involves two types of twisting, axial-type (50%) and loop-type (50%). In the axial type, the cecum twists in the axial plane without inversion, rotating about its long axis and the cecum occupies the right half of the abdomen, usually the right lower quadrant. In the loop type, the cecum twists and inverts, so that the pole of the cecum may occupy the left upper quadrant. Cecal bascule is a variant of cecal volvulus that refers to abnormal location of the dilated cecum, due to anterior folding without any torsion (9).

Plain abdominal radiography can show the typical appearance of a 'coffee-bean'-shaped dilated cecum with an air-fluid level, which may be located almost anywhere in the abdomen ([Fig 13](#)). A collapsed left colon and air- or fluid-filled dilated small bowel may also be seen. Clinical manifestations and findings on plain film in patients with cecal volvulus can be confused with gastric distension, small bowel obstruction, and sigmoid volvulus (6, 9).

CT can be useful in differentiation of cecal volvulus from its mimics, detection of the presence and location of cecal volvulus, evaluation of associated anatomic variants, and identification of fetal complications, such as ischemia and perforation. Distended ectopic cecum with a whirl sign on CT is diagnostic of cecal volvulus. The whirl sign is composed of twisted loop and mesentery containing the collapsed cecum, with adjacent bowel and the fatty mesentery containing the ileocecal vessels. The "coffee-bean" sign and the "bird-beak" sign are also common CT findings with cecal volvulus. The "coffee-bean" sign refers to the dilated cecum filled with air and fluid with haustral creases. The "bird-beak" sign is a description of tapering efferent and afferent bowel loops at the point of volvulus ([Fig 13](#)) (6, 9).
**Fig.**: Figure 13. Cecal volvulus in a 18-year-old man with abdominal distention. (a) Abdominal radiograph shows air-distended cecum in the coffee-bean shape (*) in the left abdomen.

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Fig.: Figure 13. Cecal volvulus in a 18-year-old man with abdominal distention. (b) Axial contrast-enhanced CT shows dilated cecum (*) in the left abdomen and the whirl sign (arrows).

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Fig.: Figure 13. Cecal volvulus in a 18-year-old man with abdominal distention. (c) Coronal reformatted contrast-enhanced CT shows dilated cecum (*) with beak-like tapering (arrow) in the left abdomen. Note the reversal of the normal relationship between superior mesenteric artery (black arrowhead) and superior mesenteric vein (white arrowhead).

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
**Fig.**: Figure 13. Cecal volvulus in a 18-year-old man with abdominal distention. (d) Intraoperative photograph shows cecal volvulus.

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

**Sigmoid volvulus** is also an abdominal emergency and should be differentiated from cecal volvulus. It is generally considered to be an acquired condition. Most commonly, patients with sigmoid volvulus have chronic constipation and abdominal distension, which can be associated with a high-fiber diet, pregnancy, hospitalization or institutionalization, or a large pelvic mass. These conditions may produce a long, redundant sigmoid colon with an elongated mesentery, which is prone to twisting. Imaging features are similar to those of cecal volvulus, except that the sigmoid colon, not the cecum is involved (**Fig 14**) (6).
Fig.: Figure 14. Sigmoid volvulus in a 75-year-old man with abdominal pain. (a) Plain abdominal radiography shows an air-filled, dilated sigmoid colon (*) arising from the pelvis. Note a percutaneous endoscopic gastrostomy tube.

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
**Fig.**: Figure 14. Sigmoid volvulus in a 75-year-old man with abdominal pain. (b) Coronal reformatted contrast-enhanced CT image shows dilated sigmoid colon (*) with the beak sign (arrow).

**References**: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

**Meckel's diverticulum**

Partial or complete failure of involution of the omphalomesenteric duct results in a spectrum of anomalies, including Meckel's diverticulum, with or without an omphalomesenteric ligament (or fibrous band) connecting the umbilicus, omphalomesenteric fistula, omphalomesenteric cyst, omphalomesenteric sinus, and omphalomesenteric ligament. Meckel's diverticulum is the most frequent (98% of cases) of the omphalomesenteric duct anomalies and the most prevalent congenital malformation of the small bowel (2, 10, 11).
Although it is not based on accurate pathologic data, Meckel's diverticulum is often described according to the so-called "Rule of 2's". It is seen in approximately 2% (0.3-3%) of the population, is usually detected in the antimesenteric border of the terminal ileum within 2 feet proximal to the ileocecal valve, is about 2 inches in length, occurs about equally in males and females, although males are two times more likely to have complications, may have two types of heterotopic mucosa (stomach and pancreas), and is usually symptomatic before 2 years of age (10).

It is a true diverticulum, contains all layers of the intestinal wall, and has its own blood supply. Meckel's diverticulum is usually asymptomatic, but approximately 2-4% of cases are symptomatic, with complications such as bleeding, small bowel obstruction, inflammation, and neoplastic changes. GI bleeding is the most common complication in children with Meckel's diverticulum, whereas small bowel obstruction and diverticulitis are the main complications seen in adults. Intestinal obstruction can result from an inverted Meckel's diverticulum with or without intussusception, volvulus around fibrous bands, inflammatory adhesion, enteroliths being expelled from the diverticulum, and Littre's hernia. Diverticulitis results from ulceration, secondary to acid secretion from ectopic gastric mucosa. Additionally, obstruction of diverticulum by an enterolith or foreign body impaction can also lead to inflammation, necrosis, and eventual perforation. GI bleeding occurs often in adults, which occurs as a result of peptic ulcers of the diverticulum in the presence of heterotopic gastric mucosa. Neoplasms arising in Meckel's diverticulum, such as carcinoid tumors, GIST, and leiomyoma, are very rare. The most frequent neoplasm is a carcinoid tumor (2, 10, 11).

Meckel's diverticulum may appear as a fluid- or gas-filled blind-ending pouch, arising from the antimesenteric side of distal ileum on CT. However, CT has poor sensitivity for the detection of Meckel's diverticulum in uncomplicated cases, because the appearance of Meckel's diverticulum usually resembles a normal loop of bowel. Thus, the diagnosis can be suggested if features of known complications of Meckel's diverticulum can be detected on CT. These features are most detected around the small bowel of the right lower quadrant or mid-lower abdomen.

The appearance of Meckel's diverticulum on CT will vary according to the complications (2, 11). Typical CT findings of intestinal obstruction complicated with Meckel's diverticulum are isolated small bowel obstruction, or intussusceptions with small bowel obstruction. Inverted Meckel's diverticulum that serves as a lead point for intussusceptions appears as a central area of fat attenuation surrounded by a thick collar of soft-tissue attenuation on CT, which results pathologically from entrapment of mesenteric fat within the inverted diverticular sac (Fig 15). CT findings of volvulus from fibrous bands associated with small bowel obstruction are similar to those of isolated
small bowel obstruction. Thus, it is generally difficult to identify a case of obstruction by CT in a patient with volvulus from fibrous bands or inflammatory adhesion (2, 11).

**Fig.**: Figure 15. Inverted Meckel's diverticulum with intussusception in a 28-year-old man with intermittent abdominal pain. (a) Volume-rendering three-dimensional contrast-enhanced CT shows an intraluminal mass of fat attenuation (arrow) protruding into the ileal lumen.

**References**: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
**Fig.**: Figure 15. Inverted Meckel's diverticulum with intussusception in a 28-year-old man with intermittent abdominal pain. (b) Photograph of the surgical gross specimen shows an inverted Meckel's diverticulum.

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

Meckel's diverticulitis appears as an inflammatory cystic mass attached to adjacent small bowel on CT (**Fig 16**). Inflammatory changes in the surrounding mesentery and adjacent fluid collection are also helpful features in diagnosing diverticulitis. Calcifications related to enteroliths are occasionally present within the inflamed diverticulum. When active bleeding occurs at the time of CT scanning, luminal extravasation of contrast material can be present (**Fig 17**).
**Fig.**: Figure 16. Perforated Meckel's diverticulitis in a 28-year-old woman with lower quadrant pain. (a) Ultrasound shows a thick-walled, blind-ending tubular structure with gut signature (*). Note a focal wall defect (arrowhead), indicating a peptic ulcer in the heterotopic gastric mucosa.

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
**Fig.**: Figure 16. Perforated Meckel's diverticulitis in a 28-year-old woman with lower quadrant pain. (b) Contrast-enhanced CT shows a thick-walled, blind-ending tubular structure (*) with a focal wall defect (arrowhead) located at midline pelvis. Note adjacent fluid collection (short arrow) and extraluminal gas (long arrow). At pathology, perforated Meckel's diverticulitis with heterotopic gastric mucosa was confirmed.

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Figure 17. Meckel's diverticulum in a 16-year-old man with hematochezia for 3 days. (a) Non enhanced CT image shows the fluid-fluid level that forms a lower layer of high attenuation in a blind-ending pouching bowel-like structure (arrow) from the antimesenteric border of the terminal ileum in the left lower quadrant.

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Fig.: Figure 17. Meckel's diverticulum in a 16-year-old man with hematochezia for 3 days. (b) Contrast-enhanced CT image obtained in arterial phase show no active extravasation within the Meckel's diverticulum (arrow). Note extravasated contrast media (arrowheads in a and b) in the lumen of terminal ileum due to previous angiography at another hospital.

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

Neoplasms or heterotopic tissues in Meckel's diverticulum may appear as nodular or polyploid intraluminal masses (*Fig 18*) (2, 11).
Fig.: Figure 18. Heterotopic gastric tissue in the Meckel's diverticulum of a 20-year-old man. Contrast-enhanced CT shows a 2-cm, well-defined, hypervascular intraluminal mass-like lesion (arrow) in the blind-ending tubular structure (*). Pathology confirmed the presence of ectopic gastric tissue in Meckel's diverticulum.

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

Hirschsprung disease and hypoganglionosis

Hirschsprung disease (HD) is a congenital condition and is characterized by complete or partial obstruction secondary to the total absence of intramural ganglion cells in the Meissner (submucosal) and Auerbach (myenteric) neural plexuses, principally affecting the rectosigmoid or rectal segments. It results from defective migration of the ganglion cell precursors to the hindgut during the 5th to 12th weeks of gestation (12). Hypoganglionosis (HG) is defined as a significantly small number of intramural ganglion
cells in the intestine. The cause of HG remains unclear. However, HG can be classified into congenital and acquired forms. Congenital HG is consistent with the hypogenesis of the ganglion cells, and acquired HG is consistent with degeneration and a decrease in the number of ganglion cells, due to ischemia, intramural inflammation, and/or infectious disease (13).

HD and HG usually manifest before the age of 5 years. Rarely, the diseases are initially diagnosed in adults, who have milder forms of the disease and go undiagnosed early in their lives (12, 13). The diagnosis of HD or HG is usually much more difficult in adults than in infancy. This is because of the rarity of the disease in adults and the higher incidence of short or ultrashort segment aganglionosis with relatively mild symptoms during the early stage of the disease. The correct diagnosis is based on barium enema, anorectal manometry, and most importantly, a full-thickness rectal biopsy (12).

The most reliable finding with the barium enema is a transitional zone between the aganglionic or hypoganglionic distal segment and the dilated proximal colon with normal ganglion cells. The proximal colonic segment is more dilated because the aganglionic segments showing HD is more contracted, compared with the rectum showing HG. CT can depict the correct transition zone, and also rule out other obstructive causes, such as rectal cancer (Fig 19).
Fig.: Figure 19. Congenital megacolon in a 32-year-old woman with a long-history of constipation. Sagittal reformatted contrast-enhanced CT shows markedly dilated feces-filled proximal upper part of rectum with a transition zone (arrows) and a distal narrowed rectum. Pathology confirmed aganglionic segments.

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

Thus, adult HD or HG should be considered on the basis of the CT finding of a marked colonic dilatation containing a large amount of solid feces without any other obstructive lesion at the transitional zone, and narrowed distal colorectal segment in adult patients with history of long-standing constipation from childhood (12).

Enteric duplication cyst

Enteric duplication cyst is a rare congenital anomaly that may occur anywhere along the GI tract on the mesenteric side. Duplication cyst is most common in the small bowel, particularly the distal ileum, followed by the esophagus, stomach, and colon. Duplication
cyst is usually attached to the GI tract, has smooth muscle in its wall, and is lined with GI epithelium. The etiology of enteric duplication remains unclear. However, an incomplete merging of vacuoles, the abnormal recanalization hypothesis, is a popular idea. During the 6th week of development, proliferation of epithelium completely occludes the lumen of the gut, and then vacuoles develop within the occluded lumen. Normally, vacuoles coalesce until the lumen of the gut tube is fully recanalized (Fig 20).

![Diagram of gut development](image)

**Fig.**: Figure 20. Formation of the definite gut lumen.

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

Duplication cyst can develop from abnormal persistence of vacuoles. Other hypotheses include persistent embryologic diverticular theory, bronchopulmonary-foregut malformation, intrauterine vascular accident theory, and the abortive twinning theory (14).

There are two general types: cystic duplication accounts for two-thirds of cases, is spherical in shape, and has no communication to the bowel lumen, and tubular
duplication that accounts for one-third of cases and communicates directly with the bowel lumen. Most enteric duplications present during the first year of life. Cystic duplication occasionally manifests in adults and is usually found incidentally on endoscopy or imaging studies, because it infrequently causes symptoms. Complications are rare, but may include obstruction by volvulus or intussusception, bleeding, infection, or perforation. GI bleeding may occur as a result of peptic ulcers in the presence of heterotopic gastric mucosa. Malignant changes from duplication cyst are extremely rare. Surgical excision is the treatment of choice, as malignant tumors may develop from the mucosa (14).

Ultrasound plays a critical role in the evaluation of duplication cysts. The characteristic US appearance consists of an echogenic inner mucosa surrounded by a hypoechoic outer muscular layer, which presents as a double-wall or muscular-rim sign. This sign is often found in over 50% of patients. CT is not the usual method for the evaluation of duplication, but CT can show the location and extent of the cyst, as well as complications and associated other anomalies. On CT, a duplication cyst manifests as a fluid-filled cystic mass with a thick, slightly enhancing wall arising from the GI wall or extrinsic to the GI wall (Fig 21). Increased attenuation within cysts may be due to hemorrhage or proteinaceous materials. A cystic mass with thick enhancing wall or septa and surrounding inflammation may indicate a duplication cyst complicated by infection. Enhancing solid foci within the cyst may suggest a malignant change (Fig 21) (14).
Fig.: Figure 21. Collision tumor arising from gastric duplication cyst in a 41-year-old woman with dyspepsia. (a) Axial contrast enhanced CT shows a well-defined gastric subepithelial cystic lesion (*) widening of the gastric wall between the gastric mucosal and serosal layer. Note gastric mucosa (arrowheads).

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
**Fig.:** Figure 21. Collision tumor arising from gastric duplication cyst in a 41-year-old woman with dyspepsia. (b, c) Coronal reformatted contrast-enhanced CT image (b) and coronal T2 weighted RARE image (c) show a gastric subepithelial cystic lesion with invasion to the pancreas. Note thick septa or wall (arrows) in the inferior portion of the cystic mass.

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
**Fig.** Figure 21. Collision tumor arising from gastric duplication cyst in a 41-year-old woman with dyspepsia. (b, c) Coronal reformatted contrast-enhanced CT image (b) and coronal T2 weighted RARE image (c) show a gastric subepithelial cystic lesion with invasion to the pancreas. Note thick septa or wall (arrows) in the inferior portion of the cystic mass.

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Fig.: Figure 21. Collision tumor arising from gastric duplication cyst in a 41-year-old woman with dyspepsia. (d) EUS image shows an intramural subepithelial cystic mass which has fluid-fluid level (*) and intracystic solid portions (arrow) with direct invasion to pancreas (P). Intracystic solid portion may suggest malignant transformation.

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
Fig.: Figure 21. Collision tumor arising from gastric duplication cyst in a 41-year-old woman with dyspepsia. (e) Photograph of the surgical gross specimen shows a gastric duplication cyst (arrowhead) with intracystic and pericystic solid components (arrows), which invades the pancreatic tail (P).

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of
**Fig.:** Figure 21. Collision tumor arising from gastric duplication cyst in a 41-year-old woman with dyspepsia. (f) Photomicrograph (original magnification, ×200; hematoxylin and eosin staining) shows collision tumor of gastric duplication containing both adenocarcinoma (left) and squamous cell carcinoma (right).

**References:** N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

**Miscellaneous**

**Internal hernias** are defined as the herniation of a viscus through defects, pouches, or openings in the abdominal cavity. Although acquired defects of the mesentery or omentum due to previous surgery or trauma are more common causes of internal hernia in adults, opening or pouches caused by congenital anomalies of intestinal rotation and peritoneal fixation, such as paraduodenal hernia, paracecal hernia, intersigmoid hernia, and transmesenteric hernia, can also be underlying conditions favoring hernia.
Rarely, normal openings, such as the foramen Winslow, can lead to internal hernias. Paraduodenal hernia is the most common type of all internal hernias and is caused by small bowel entrapment under the right or left mesocolon. CT findings of internal hernia include distended bowel loops with abnormal locations and clustered small bowel loops due to encapsulation within the hernia sac (Fig 22).

Fig.: Figure 22. Left paraduodenal hernia in a 51-year-old man with abdominal pain. Contrast-enhanced CT shows a sac-like bowel loop (arrows) in the left paraduodenal fossa. Note anterolaterally displaced inferior mesenteric vein (arrowhead).

References: N. K. Lee; Department of Radiology, Pusan National University Hospital, Busan, KOREA, Republic of

Additional findings, such as mesenteric vessel engorgement, crowding, angulation, or stretching, are commonly seen. Knowledge of the normal anatomy of the peritoneal cavity, such as hallow visscus, peritoneal reflection, and the mesenteric or colic vessels, and the characteristic location of each type of internal hernia are mandatory to identify the abnormal location of herniated bowel loops in patients with internal hernia on CT (15).
Conclusion

Most congenital anomalies of the GI tract present in newborns and young infants. However, some patients will present years later or remain asymptomatic for life. Adult presentation of congenital anomalies in the GI tract can often be mistaken for other common abdominal diseases, because these conditions often manifest chronic and nonspecific symptoms and the incidence of congenital anomalies in the GI tract progressively decreases in the older population. Thus, radiologic examinations play a useful role in the evaluation of congenital anomalies of the GI tract in adults. Because of the widespread use of CT for evaluation of adult patients suspected of having nonspecific abdominal symptoms or acute abdomen, various congenital anomalies of the GI tract are increasingly being discovered by CT in adults. Identification of a variety of symptomatic congenital anomalies in the GI tract by CT can be important in establishing the correct diagnosis and determining appropriate treatment.
Personal Information

References


