Radiological characterization of congenital upper gastrointestinal tract abnormalities in children

Award: Magna Cum Laude
Poster No.: C-1418
Congress: ECR 2015
Type: Educational Exhibit
Authors: A. Coma, G. Enriquez, I. Barber, A. Castellote, J. Piqueras, E. Vazquez; Barcelona/ES
Keywords: Barium meal, Contrast agent-oral, Fluoroscopy, Digital radiography, Ultrasound, Gastrointestinal tract, Abdomen, Paediatric, Congenital, Obstruction / Occlusion
DOI: 10.1594/ecr2015/C-1418

Any information contained in this pdf file is automatically generated from digital material submitted to EPOS by third parties in the form of scientific presentations. References to any names, marks, products, or services of third parties or hypertext links to third-party sites or information are provided solely as a convenience to you and do not in any way constitute or imply ECR’s endorsement, sponsorship or recommendation of the third party, information, product or service. ECR is not responsible for the content of these pages and does not make any representations regarding the content or accuracy of material in this file.

As per copyright regulations, any unauthorised use of the material or parts thereof as well as commercial reproduction or multiple distribution by any traditional or electronically based reproduction/publication method is strictly prohibited.

You agree to defend, indemnify, and hold ECR harmless from and against any and all claims, damages, costs, and expenses, including attorneys’ fees, arising from or related to your use of these pages.

Please note: Links to movies, ppt slideshows and any other multimedia files are not available in the pdf version of presentations.

www.myESR.org
Learning objectives

• To review common and uncommon congenital lesions affecting the upper gastrointestinal tract in children.
• To illustrate the radiologic features of these conditions using various imaging techniques.
• To provide tips to avoid potential diagnostic errors.
Background

Congenital abnormalities of the upper gastrointestinal tract (GI) are relatively common in children and present in a wide range of clinical settings. In most patients, the clinical symptoms appear at birth, whereas in others, the manifestations appear later. They may be an incidental finding or present with an acute onset in neonates, infants, older children, and even in adults.

Radiological examination is essential to suggest the diagnosis. Although abdominal radiography and contrast gastrointestinal examination can provide the diagnosis, particularly in neonates, sonography has come to play a significant role in children. However, an exquisite imaging method and technique are essential. Computed Tomography CT and Magnetic resonance imaging (MRI) may be helpful in selected cases.

The clinical presentation, imaging features, and diagnostic approach for congenital upper GI tract abnormalities are discussed. Associated abnormalities are reviewed.
Findings and procedure details

COMMON CONGENITAL UPPER GASTROINTESTINAL ABNORMALITIES

GASTROINTESTINAL ATRESIA

Gastrointestinal atresia is a congenital absence or abnormal narrowing of the alimentary tract. It is thought to be the result of an imperforated web or diaphragm.

After birth, the most common clinical setting in intestinal atresia is neonatal obstruction.

Associated anomalies include malrotation, volvulus, and gastroschisis. Ileal atresia is associated with cystic fibrosis in up to 12% of patients.

Surgical repair is the treatment of choice.

Duodenal, jejunal, and proximal ileal atresia are quite common. Pyloric atresia is an uncommon upper gastrointestinal abnormality.

Imaging findings

Intestinal atresia is usually diagnosed prenatally based on signs of fluid-filled dilated bowel loops on prenatal ultrasound (US). Intestinal hyperrefringency at prenatal US also suggests the diagnosis. Fluid signal intensity in dilated bowel loops may be seen on prenatal T2-WI MRI sequences.

Postnatal radiography is often diagnostic and determines whether an obstruction is present. One, two, three, or more air-filled dilated bowel loops (depending on the site of the atresia) with no distal bowel gas are typical findings on radiography. Evaluation of distal bowel gas may be challenging. Three hours after birth, the entire small bowel is usually filled with gas, and 8 to 9 hours after birth, there is gas in the sigmoid colon (Fig. 1).

However, there may be an absence of abdominal gas in neonates undergoing mechanical ventilation or nasogastric aspiration, and passage of gas may be delayed when a neonate experiences a traumatic injury, septicemia, hypoglycemia, or brain damage.

DUODENAL ATRESIA

Duodenal atresia is the most common type of small intestinal atresia.
Antenatal US findings include polyhydramnios, and dilatation of the stomach and proximal duodenum. The **classic postnatal radiography** presentation of duodenal atresia is that of a "double bubble", where the stomach and duodenum are distended (Fig. 2). Distal bowel gas is usually absent. Nonetheless, the imaging findings vary, and the neonate may show a single bubble or, less commonly, distal bowel gas due to anomalous biliary ductal anatomy.

**JEJUNAL AND PROXIMAL ILEAL ATRESIA**

The **classic imaging presentation** of jejunal atresia is that of a "triple bubble" where 3 dilated segments jut out from the remaining gasless loops (Fig. 3).

Obstructions occurring above the mid-ileum are referred to as high obstructions, whereas those involving the distal ileum and colon are low obstructions. This distinction is mandatory: In high gastrointestinal obstruction, radiography often suffices for the diagnosis, but in low intestinal obstruction, contrast enema examination is needed to provide a specific diagnosis and guide adequate therapy. Ileal atresia may be included in high or low intestinal obstruction, depending on the site of the atresia. Differentiating between jejunal or proximal ileal atresia and distal ileal or colon atresia may be difficult, and **contrast enema examination is required in some cases**. When the atresia is in a high bowel segment, the colon caliber will be normal in size as the remaining distal bowel loops produce sufficient secretions. When atresia occurs in low bowel segments, the colon caliber is decreased (functional microcolon).

**MECKEL'S DIVERTICULUM**

Meckel's diverticulum is a true diverticulum that contains all the intestinal layers and has its own vascularization. Meckel's diverticulum forms when the omphalomesenteric duct fails to involute. It is usually asymptomatic. In symptomatic children, bleeding and small bowel obstructions are the most common symptoms. The small bowel obstruction can be caused by intussusception (Fig. 4 and 5), an inverted diverticulum, fibrous bands and volvulus, or inflammatory adhesions (Fig. 6). Neoplasms have also been described.

**Imaging findings** depend on the clinical setting and the type of complication. Asymptomatic Meckel's diverticulum is not usually seen at imaging. On **US or CT**, a small fluid-filled or air-filled lesion may be seen in the antimesenteric region of the terminal ileum. As the diverticulum usually contains gastric mucosa, the most specific test for Meckel diverticulum is a 99mTc pertechnetate scan (Fig. 7). Heterotopic gastric tissue within the diverticulum may appear as a hypervascular nodular lesion on contrast-enhanced CT. Inverted Meckel's diverticulum that serves as a lead point for intussusception appears as a central area of entrapped mesenteric fat surrounded by soft tissue (Fig. 4 and 5).
INTESTINAL MALROTATION

Disorders of intestinal rotation and fixation are commonly referred to as *malrotation*. Diagnostic imaging of intestinal malrotation is challenging but extremely important, as malrotation predisposes to an acute life-threatening condition: volvulus.

The origin:

Anatomical development of the intestinal tract is a complex process. Malrotation should be understood as a continuum of abnormalities reflecting a failure in rotation or fixation at any stage in midgut development. In brief:

- **Early rotation failure**: After the first 90° rotation, the duodenum lies to the right of the superior mesenteric artery and the distal colon to the left. There is no further rotation, and the small bowel is located on the right and the colon on the left. Small bowel location on the right with a normally located colon, or normal duodenal position with nonrotation of the colon, although rare, have also been described.
- **Failure of rotation occurring during the final 180° counter-clockwise rotation**: The abnormally rotated bowel does not develop a normal mesenteric attachment.

Failure of small bowel and colon fixation may lead to additional abnormalities, such as undescended cecum or mobile cecum. The cecal fixation process terminates in the first months after birth; consequently, the incidence of cecal malfixation decreases with age. When present in older children, it may predispose to volvulus or intussusception (Waugh’s syndrome).

**Ladd’s bands** are peritoneal bands that attempt to attach the malpositioned bowel. The bands run from the cecum and proximal colon to the right upper quadrant retroperitoneum, often entrapping the duodenum.

The many faces of malrotation

Malrotation may be an incidental finding in an asymptomatic patient, an imaging finding when studying chronic abdominal symptoms in a child, or a surgical emergency when there is associated malrotation and volvulus.

Chronic symptoms of malrotation include chronic intermittent pain, intermittent vomiting, diarrhea, constipation, and a history of acute abdominal pain. Bowel obstruction with bilious vomiting is the classic clinical presentation. Hematochezia due to bowel ischemia may occur in some cases.
Malrotation is present in most patients with diaphragmatic hernia, gastroschisis and omphalocele. It is also seen in the majority of patients with heterotaxy syndromes. Consequently, when heterotaxy syndrome is recognized, further workup is needed to exclude intestinal malrotation (Fig. 8 and 9). Other associated anomalies of malrotation include chromosomal anomalies (Down syndrome, the most frequent), intestinal atresia, cloacal extrophy and prune belly syndrome, megacystis microcolon intestinal hypoperistalsis syndrome, intestinal neuronal dysplasia, annular pancreas and rarely, Hirschsprung disease.

The imaging battle

**Radiography** is generally the first imaging technique used, and it rarely suggests the diagnosis of malrotation. However, radiography is essential to exclude other conditions, especially in a non-bilious vomiting neonate. It may also guide further imaging investigation. The most common radiographic finding in malrotation is a normal bowel gas pattern (Fig. 10). Small bowel on the right and colon on the left, seen in some cases, suggest the diagnosis (Fig. 13). When malrotation with volvulus or a peritoneal obstructing band are present, the "double bubble" appearance may simulate duodenal atresia.

**Contrast gastrointestinal examination** is the reference standard imaging method. A true and slight right lateral view should be included when studying the second, third, and forth portion of the duodenum, which are fixed to the retroperitoneum and posterior in location. When contrast reaches the distal portion of the duodenum, the duodenojejunal junction is documented in the anteroposterior view.

Signs of malrotation include abnormal position of the duodenojejunal junction, spiral, "corkscrew" or Z-shaped course of the distal duodenum and proximal jejunum, and location of the proximal jejunum in the right abdomen (Fig. 14 and 15). When volvulus is present, the "corkscrew" appearance adds a narrowed lumen with distal beaked appearance and proximal dilated loops.

Normal variants such as jejunum in the right upper quadrant as the sole abnormality, duodenojejunal junction over the left pedicle in the anteroposterior view and mobile duodenum may be confusing. Normal contrast upper GI examinations have also been described in malrotation. Indeterminate findings or a high suspicion of malrotation with a normal upper GI appearance warrants further workup. The position of the cecum should be demonstrated by performing an enema (Fig. 10, 11 and 12) or following barium through the small bowel (Fig. 14 and 15).

**US** has come to play an important role in evaluating gastrointestinal abnormalities in children. High-frequency transducers and an exquisite imaging method and technique are essential. When malrotation is suspected and upper gastrointestinal contrast study
is indeterminate or equivocal, US can add useful information. The relationship between the superior mesenteric vein and artery should be determined. Normally, the superior mesenteric vein is to the right of the artery (Fig. 16). The 3rd portion of the duodenum should be seen crossing (Fig. 17). Recent studies have delineate duodenal anatomy on US using water instillation. The ileocecal valve can also be depicted by US with a meticulous technique.

When volvulus is present, US and CT may show the superior mesenteric vein to the right, coursing around the superior mesenteric artery (whirlpool sign) (Fig. 18). Signs of bowel ischemia should be sought. Dilated bowel loops, thickened bowel walls, and aperistaltic loops suggest ischemic intestinal injury on US. Thin walls and absent Doppler signal are unusual, although highly suggestive features. CT is not routinely performed in children to avoid radiation exposure.

**CT and MRI** show reversed positioning of the superior mesenteric vein and artery, a duodenum that does not cross midline and small bowel on the right. The diagnosis is usually made in a patient undergoing CT or MRI, in whom the condition is not suspected, or as an incidental finding (Fig. 11, 19).

**The end**

Laparotomy or a laparoscopic Ladd procedure is the surgical technique of choice. It consists of laparotomy with reduction of the midgut volvulus, division of peritoneal bands, placement of the small and large bowel in a state of nonrotation and appendectomy. After treatment, patient care should continue. Although recurrent volvulus is rare, it can occur (Fig. 19 and 20).

**UNCOMMON UPPER GASTROINTESTINAL CONGENITAL ABNORMALITIES**

**GASTROINTESTINAL DUPLICATION CYST**

Gastrointestinal duplication cysts are spherical or tubular lesions lined by gastric or enteric epithelium and surrounded by a layer of smooth muscle (Fig. 21). They can arise anywhere from the esophagus to the rectum, and be attached to these structures or located nearby. The most common locations are the distal ileum and esophagus. Gastric and duodenal duplications are the least common. The greater curvature of the stomach is the most frequent location of gastric duplication cysts. Rarer locations include the pleura, retroperitoneum, pancreas, and liver.
Clinical aspects predominantly depend on the size of the cyst. Typically, the lesions are asymptomatic or manifest with abdominal pain, vomiting, or a palpable mass. As they are lined with alimentary tract epithelium, they can manifest with bleeding (Fig. 22) or rupture with peritonitis. Patients with intestinal duplication cyst may also present with intestinal obstruction or intussusception (Fig. 23). Associated small bowel atresia has been described.

Imaging findings

US is the imaging modality of choice. Prenatal examination may confuse a gastric duplication cyst with duodenal atresia, as a "double bubble" sign may be visualized. The typical US findings are an inner hyperechoic mucosa and outer hypoechoic muscular wall (Fig. 21 and 22). Peristaltic activity, if present, can confirm the diagnosis. The typical image is not always seen, and a double wall sign may be present in other cystic lesions, such as Meckel's diverticulum. Enteric duplication cyst may contain ectopic gastric mucosa and be detected with technetium-99m pertechnetate study.

CT and MR can be used to determine the location, extension, and nature of the cyst (Fig. 24).

ANNULAR PANCREAS

Annular pancreas may cause neonatal duodenal obstruction or be asymptomatic, with discovery as an incidental finding in adulthood. Pancreatitis, ulceration, and ductal obstruction have also been described. In annular pancreas, a band of pancreatic tissue connected with the pancreatic head surrounds the second duodenal portion.

Associated anomalies, reported in up to 70% of patients, include duodenal atresia, Down's syndrome, tracheoesophageal fistula, congenital heart defects, pancreas divisum, and heterotaxy syndrome.

Imaging findings

When annular pancreas causes duodenal obstruction, radiography shows the classic "double bubble" sign seen in duodenal atresia. Distal bowel gas is often present (Fig. 25), whereas in complete obstruction, it may be absent. Contrast GI examination, the second imaging tool used to investigate obstruction, shows duodenal stenosis and extrinsic circumferential narrowing of the descending duodenum (Fig. 26 and 27). It is difficult to demonstrate pancreatic tissue around the duodenum on US examination due to gas in distended bowel loops. However, an anomalous pancreatic beak can suggest the diagnosis. CT and MRI depict normal pancreatic tissue encircling the duodenum. MR cholangiopancreatography and endoscopic retrograde cholangiopancreatography
(ERCP) are the imaging modalities of choice when evaluating pancreas anatomy. However, these studies are not routinely performed.

**ANTRAL AND DUODENAL WEB**

Mucosal diaphragms are rare, often misdiagnosed, congenital disorders that cause complete or partial upper gastroduodenal obstruction.

Antral and duodenal webs have been described in association with heterotaxy syndrome.

**ANTRAL WEB**

Antral web causes complete or partial gastric outlet obstruction. **Radiography** may show gastric dilatation (Fig. 28). **US** is often performed to rule out HPE. At US, persistent echogenic structures are seen extending centrally from the lesser and greater curvatures in the prepyloric region (Fig. 29). **Contrast GI examination** findings in antral web are normal pylorus, proximal gastric dilatation, gastric emptying delay, and an intraluminal smooth web (Fig. 30). Endoscopic resection is the treatment of choice (Fig. 31).

**DUODENAL WEB**

Duodenal web occurs in the ampulla of Vater region. The common bile duct is incorporated inside the web and may drain above or below it. An echogenic linear structure may be depicted on **US** or a smooth intraluminal linear structure on **contrast GI examination**. The different types of duodenal webs include duodenal atresia (imperforate web), imperforate intraluminal duodenal diverticulum (windsock web), and perforated duodenal web (Fig. 32). Treatment options include resection of the web or duodenoduodenostomy when obstruction is complete.

**PYLORIC ATRESIA**

Pyloric atresia accounts for only 1% of intestinal atresia cases. It presents as outlet gastric obstruction, but it has an excellent prognosis with adequate surgical treatment. It is often associated with **epidermolysis bullosa** and hereditary multiple atresia.

Prenatal images show polyhydramnios and persistent gastric and esophageal dilatation without duodenal distension. When pyloric atresia is associated with epidermolysis bullosa, **US** may show echogenic particles within the amniotic fluid. Postnatal images may show a "single bubble" corresponding to distended stomach on **radiography** and anomalous pylorus morphology on **US**.

**HETEROTOPIC PANCREAS**
Ectopic remnants of pancreatic tissue can appear in various locations, usually in the submucosa of the gastric antrum or the proximal portion of the duodenum. Patients are usually asymptomatic, but sometimes the ectopic pancreatic tissue is functional and inflammation occurs. Obstruction and intussusception may take place when the location is the small bowel. Malignant transformation has also been described. **Contrast GI examination** depicts a smooth submucosal lesion, which may present central umbilication. Laparoscopic resection is the treatment of choice.

**AGANGLIONOSIS**

Aganglionosis is an uncommon congenital abnormality that is easily confused with meconium ileus or intestinal atresia. Aganglionosis results from abnormal development of the enteric nervous system, and can affect the jejunum, ileum, or colon. Newborns present with signs and symptoms of bowel obstruction, and dilated bowel loops are seen on **radiography** (Fig. 33). **Contrast GI examination** is the image technique of choice. It suggests the diagnosis and excludes other entities (Fig 34 and 35).

**INTERNAL HERNIA DUE TO A CONGENITAL DEFECT**

Internal hernias often result from bowel fixation failure or incomplete mesentery fusion. The most common internal hernias in children are transmesenteric hernia, caused by a small bowel mesenteric defect near the ileocecal region or the angle of Treitz.

Common abnormalities associated with congenital transmesenteric hernia include intestinal atresia, malrotation, and peritoneal band.

Imaging plays an important role in the diagnosis. Abdominal radiography and US are limited for identifying an internal hernia, but they are useful for detecting bowel obstruction and ruling out other entities. Contrast GI studies and CT, although challenging, may suggest the diagnosis. On **contrast GI examination**, encapsulated, clustered, or dilated bowel loops are seen in an anomalous location, and a herniated sac is delineated in some cases. Anomalous peristalsis is also seen. **CT** can demonstrate the obstruction point and depict anomalous relationships between the mesenteric vessels, which may be twisted and stretched, usually accompanied by venous or lymphatic engorgement.

**MECONIUM PERITONITIS**

Meconium peritonitis is a chemical peritonitis cause by an intrauterine bowel perforation. It may be idiopathic or have an underlying cause, such as intestinal atresia, intussusception, meconium ileus, or volvulus. Meconium produces an intense peritoneal inflammatory reaction. Postnatal **radiography** shows ascitis and occasionally depicts calcifications (Fig. 36) **US** signs include ascitis with echogenic foci (snowstorm appearance), encapsulated meconium pseudocysts with hyperechogenic
walls, collapsed and distended bowel loops, and peritoneal calcifications. The calcifications may extend to the scrotum through the processus vaginalis (Fig. 37). The inflammatory tissue often resolves spontaneously and peritoneal calcifications are seen postnatally (Fig. 38).
Fig. 1: Normal bowel gas pattern in a term newborn 8 hours after birth. Differentiation between colon and small bowel may be impossible, as in this case. Size and distribution can help.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
**Fig. 2:** Duodenal atresia. Radiography 10 hours after birth in a premature neonate with abdominal distension and emesis shows the classic "double bubble" sign. Note distension of stomach (asterisk) and duodenum (arrowhead). Distal bowel gas is absent, indicative of complete obstruction and duodenal atresia. Duodenal atresia may be associated with other congenital anomalies, such as anomalous biliary duct anatomy, cardiac anomalies, VACTERL (vertebral, anorectal, tracheoesophageal, renal, and limb anomalies), alveolar capillary dysplasia, and chromosomal anomalies (e.g., trisomy 21). Thirty percent of infants with duodenal atresia have Down syndrome. Hence, when duodenal atresia is suspected, obstetricians, pediatricians, and radiologists should seek features of associated anomalies.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 3: Preterm infant with hyperrefringent bowel and suspected intestinal atresia at prenatal US. Radiography shows the "three bubble" sign with air-filled stomach (asterisk), duodenum (black arrowhead), and proximal jejunum (white arrowhead). Absent distal bowel gas. Radiographic features indicate jejunal atresia. Note abnormal disposition of proximal jejunal loops in right abdomen, suggesting malrotation. Malrotation is often associated with intestinal atresia. Laparotomy revealed type IIIa proximal jejunal atresia and intestinal malrotation. Surgical treatment included end-to-end jejunal anastomosis and Ladd's procedure.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 4: Meckel's diverticulum in an 8-year-old boy with abdominal distension and bilious vomiting. US depicts dilated bowel loops with edematous mural thickening (a). Free anechoic fluid is also seen (asterisk in a). Intussusception in the lower right quadrant (b, c). Longitudinal view shows internal lymphadenopathies (arrow in b). When intussusception is seen in a child or adolescent, intussuscepted lesions such as Meckel's diverticulum or duplication cyst should be investigated. Inverted Meckel's diverticulum serving as a lead point for intussusception appears as a central area of entrapped mesenteric fat surrounded by soft tissue. Soft tissue (black asterisk in c) surrounding mesenteric fat was visualized in the axial view (c) and Meckel's diverticulum was suspected.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
**Fig. 5:** Same patient as Fig. 4. CT demonstrates intestinal occlusion signs resulting from ileo-colic intussusception (a, b and c) and depicts the inverted diverticulum (orange arrows in b) surrounding the mesenteric fat (white arrow in b) in the upper part as the lead point of intussusception. Meckel's diverticulum in the antimesenteric border of the terminal ileum was confirmed at surgery (d).

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 6: Intestinal occlusion due to Meckel's diverticulum with inflammatory adhesions in a 5-year-old boy with vomiting, diarrhea, and abdominal pain of 3 days' duration. One day before the study, diarrhea had stopped, but abdominal pain and vomiting persisted. Coronal CT images show signs of intestinal occlusion (a, b, c and d). Severe small bowel distension with free fluid between intestinal loops (a). Correct relationship between superior mesenteric vein and artery on coronal CT with MIP reconstruction (b). Collapsed small bowel loops in the superior right quadrant and mesogastric region (c). CT depicts the obstruction point where loops change caliber (d). A small fluid-filled lesion (arrows in d) was seen at this point, and Meckel's diverticulum was suspected. Surgery confirmed the diagnosis and demonstrated inflammatory adhesion around bowel loops and Meckel's diverticulum. The patient required intestinal resection.
Fig. 7: Meckel's diverticulum. A 6-year-old boy with two episodes of hematochezia at home. No fever, diarrhea, weight loss, or joint pain. On analyses, inflammatory parameters were not elevated. US shows a 14-mm nodular structure in right lower abdominal quadrant, mimicking a bowel loop (a). Selective pain was elicited at compression. No peristaltism was observed during the examination. Mural vascularization was seen on Doppler US (b). Abdominal SPECT-CT confirmed ectopic gastric mucosa in the right lower quadrant (c). Meckel's diverticulum was surgically resected (d) and pathological analysis showed ulceration and gastric metaplasia (e).
Fig. 8: Neonate with prenatal diagnosis of complex cardiopathy and situs inversus. Abdominal radiography shows liver at the left and stomach at the right. Abnormal bowel distribution was suspected.

© Radiology, Hospital Materno-Infantil Vall d’Hebron - Barcelona/ES
Fig. 9: Same patient as Fig. 8. Contrast GI series (a, b, and c) confirms intestinal malrotation. The normal duodenum ends cephalad, near the level of the pylorus, and at the left of the L2 vertebral pedicle. Note abnormal, slightly descended position of the duodenojejunal junction on right and left lateral views (arrows in a and b) and superimposed on left vertebral pedicle on anteroposterior view (arrow in c). Highly focused attention is required when evaluating a child with dextrogastria, in which a normally located duodenum presents a mirror image of normal. Malrotation is excluded when the duodenum extends to the right pedicle and upwards.

© Radiology, Hospital Materno-Infantil Vall d’Hebron - Barcelona/ES
Fig. 10: Intestinal malrotation in a 2-year-old boy with several months’ history of intermittent vomiting causing metabolic changes. Hospital care was needed during these episodes. Radiography (a) shows normal bowel gas pattern, a common finding in malrotation. Contrast GI series depicts subtle findings: slightly descended duodenojejunal junction in lateral (b) and anteroposterior (c) views that went unnoticed at this time.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
**Fig. 11:** Same patient as Fig. 10. MRI was performed to rule out annular pancreas, which can cause partial duodenal obstruction. Pancreatic anatomy was normal, but axial T2-WI (a) showed jejunal bowel loops at the right side of the abdomen (asterisk). Contrast-enhanced fat-sat T1WI (b) depicts anomalous relationship between superior mesenteric vein and artery (arrow).

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 12: Same patient as Figs. 10 and 11. Barium enema shows abnormal cecal position (asterisk). Intestinal malrotation was diagnosed. As the duodenojejunal junction and cecum do not rotate simultaneously during development, a normal cecal position does not exclude malrotation. Consequently, barium enema should not be the only study performed.
Fig. 13: Neonate with dextrocardia and intestinal malrotation. Radiography shows small bowel on the right and colon on the left. Note central position of the liver and normal position of the stomach in this case.

© Radiology, Hospital Materno-Infantil Vall d’Hebron - Barcelona/ES
Fig. 14: Intestinal malrotation in a 2-year-old boy with cyclic vomiting. He presented hematemesis and bilious vomiting during the last episode. Radiography performed 4 hours after the episode (a) showed asymmetric bowel gas pattern with gas-filled loops at the left. At physical examination, the abdomen was soft, bowel peristalsis was present, and no palpable mass was detected. Radiography performed 12 hours after the episode (b) showed a non-specific bowel pattern. Signs and symptoms of intestinal subocclusion that resolve should alert to intestinal malrotation.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 15: Same patient as Fig 14. Contrast GI series demonstrates signs of intestinal malrotation: descended (a) and right-sided (b) duodenojejunal junction and proximal jejunal loops in the right side of the abdomen (c). As barium passed through the small intestine, a high central cecal position was depicted (d).

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 16: The anatomic relationships of mesenteric vasculature should be investigated on US if intestinal malrotation is suspected. The superior mesenteric vein is to the right of the artery. However, a correct proximal relationship does not exclude malrotation, as in some cases mesenteric twisting occurs at a slightly lower position. The entire mesentery should be examined using a meticulous US technique.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 17: The retroperitoneal position of the duodenum has been proposed as a reliable anatomical marker of normal rotation. Axial imaging techniques such as US can depict the third portion (3A) of the duodenum crossing the midline. The duodenojejunal angle may be depicted on the left side, posterior to the stomach (4A).

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
**Fig. 18:** Intestinal malrotation and volvulus in a 3-year-old boy with abdominal distension and bilious vomiting. Abdominal US shows bowel loops, mesenteric fat, and mesenteric vasculature running from right to left, and from anterior to posterior (a). This is know as the "whirlpool" sign, it is easily visualized on Doppler US, and suggests intestinal malrotation complicated by volvulus (b). Malrotation predisposes to volvulus, an acute life-threatening condition in which the intestine twists around the SMA and obstructs the bowel lumen, lymphatic and venous drainage, and in some cases, arterial supply. Note slight engorgement of mesenteric venous branches (arrows) and multiple lymphadenopathies (asterisks) in this case (c).

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 19: Intestinal malrotation in a 8-year-old boy with chronic abdominal pain. Axial CT (a) and coronal CT with MIP reconstruction (b) performed in an other medical center show jejunal loops at the right and ileal loops at the left. Note the abnormal relationship between superior mesenteric vein an artery on coronal CT with MIP reconstruction (arrow).

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 20: Same patient as Fig. 19. 24 hours after the laparoscopic Ladd procedure the patient presented abdominal distension and bilious vomiting. Coronal (a) and sagital (b and c) contrast enhanced CT demonstrate signs of intestinal occlusion: dilated stomach and bowel loops, collapsed distal bowel loops and free fluid between loops. Post-procedure pneumoperitoneum and subcutaneous air bubbles are also seen. Note the obstruction point in axial and coronal images (arrow in a and c). Although the "whirlpool sign" was not seen, volvulus was confirmed at surgery. The "whirlpool" sign may not be seen, specially if it doesn't affect proximal loops. Clinical setting and signs of occlusion at imaging suggest it.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 21: Duplication cyst. Abdominal US with high frequency transducer shows a spherical anechoic thin-walled lesion with typical double-wall sign (arrow).

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 22: A 4-month-old boy with prenatal diagnosis of gastric duplication cyst and sudden abdominal pain. Abdominal US (a, b) demonstrates the cystic lesion between the greater curvature of the stomach and the left liver lobe; it has significantly enlarged and shows echogenic content. Note the double-wall sign. Echogenic content in a duplication cyst may correspond to secretions or debris. However, sudden enlargement and echogenic content often indicate infection or bleeding. Inflammatory parameters were normal in this baby, making intracystic bleeding the more appropriate diagnosis.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 23: Ileo-colic intussusception in a 3-month-old girl with vomiting. Transverse (a) and longitudinal (b) US images show a lower right quadrant mass with the characteristic appearance of ileo-colic intussusception (alternating hyper- and hypoechoic layers). Hyperechogenic mesenteric fat and lymphadenopathies are seen within (arrow in b). A spherical thin-walled lesion with a fluid-fluid level is depicted inside the intussusception (c). Doppler signal was increased around the duplication cyst in the bowel loops, due to reactive inflammatory changes. Duplication cyst is suggested as the lead point of the intussusception.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 24: Intrapancreatic duplication cyst in a 13-year-old girl with repeated episodes of weakness and non-specific epigastric pain. No nausea, vomiting, or diarrhea. Soft abdomen with no organ enlargement or palpable masses. Laboratory tests showed mildly elevated pancreatic amylase. CA19.9 and CEA tumor markers were normal. On US study, a 3.5cm-diameter well-defined homogenic echoic lesion was demonstrated adjacent to spleen, left kidney, and pancreatic tail (a). Doppler showed no vascularity (b). Linear transducer US depicts a thin echogenic wall (c). Absent double wall sign does not exclude duplication cyst. Abdominal MR imaging discloses a lesion related to the pancreatic tail in the left upper abdomen with homogeneous hypointensity on T1WI, (d) and slightly heterogeneous hyperintensity on fat-sat T2WI (e). Low-dose unenhanced abdominal CT scanning used to guide fine-needle aspiration sampling reveals a rounded, well-defined, homogenic, slightly hypodense lesion at the pancreatic tail. There were no calcifications (f).

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 25: Annular pancreas in a 4-year-old boy with several month's history of constipation and vomiting. Radiography shows "double bubble" sign with distal bowel gas, excluding complete occlusion. Descending duodenum compression and annular pancreas were suspected. The differential diagnosis includes other causes of duodenal obstruction such as peritoneal fibrous band and duodenal web.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 26: Same patient as Fig. 25. Contrast GI series (a,b) shows obstructive dilatation of the stomach and proximal duodenum. Antiperistaltic contractions were noted and only small amounts of contrast material passed distally. Extrinsic compression was suggested. Note the extrinsic mass effect on the second portion of the duodenum (curved line).

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
**Fig. 27:** Same patient as Figs. 25 and 26. Surgery revealed duodenal obstruction due to an annular pancreas encircling the descending duodenum (a,b). Duodenojejunal junction was correctly located. Duodenoduodenostomy was performed, with no complications (c).

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
**Fig. 28:** Antral web in a 1-month-5-day-old infant with non-bilious vomiting and hypotonia of 24 hours' duration. Radiography shows stomach distension without associated duodenal distension: the "single bubble" sign. There is distal bowel gas. Hypertrophic pyloric stenosis would be a consistent diagnosis, but findings on US and contrast GI series ruled this out.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 29: Same patient as Fig. 28. Abdominal US depicts large gastric contractions (a), normal anatomy of pancreatic head and duodenum (b), and persistent echogenic structures (arrows in c and calipers in d) narrowing the prepyloric region lumen (c, d).

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 30: Same patient as Figs. 28 and 29. Contrast GI series shows large gastric contractions (a), normal pyloric anatomy (b), and a thin membrane in the prepyloric region (c, d).

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 31: Same patient as Figs. 28, 29 and 30. Endoscopic resection. Antral web was confirmed and resected.

© Radiology, Hospital Materno-Infantil Vall d’Hebron - Barcelona/ES
Fig. 32: Duodenal web in a 1-month-old infant with bilious vomiting of 24 hours. "Double bubble" sign is seen on abdominal US (a). Duodenal occlusion is suspected. Pancreatic anatomy appears normal on US, but contrast GI series (a,b and c) shows obstructive dilatation of the stomach and proximal duodenum, and only small amounts of contrast material passing distally. No extrinsic compression was demonstrated in this case. Duodenal web was diagnosed at surgery.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 33: Neonate with a single dilated bowel loop on prenatal US. Radiography shows a non-specific bowel loop pattern with one dilated air-filled loop. The differential diagnosis includes bowel atresia, meconium ileus, communicating duplication cyst, and aganglionosis.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 34: Same patient as Fig. 33. Enema demonstrates normal colon caliber and rules out distal ileal atresia. There were no meconium "pebbles" suggesting meconium ileus. A large contrast-filled small bowel loop and aganglionosis were suggested (asterisk in a and b). Surgical resection and pathological analysis diagnosed a fusiform communicating duplication cyst.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 35: Neonate with a single dilated bowel loop at prenatal US. Radiography (a) shows non-specific bowel loop pattern with one dilated air-filled loop in the upper abdomen. The differential diagnosis includes communicating duplication cyst and aganglionosis. Contrast GI series showed correct feeding of small bowel and colon. The lesions had a peripheral rim of contrast (arrow). A communicating duplication cyst was diagnosed.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
**Fig. 36:** Meconium peritonitis in a neonate with abdominal distension. Radiography suggests ascitis with bowel loops centrally located and depicts peritoneal calcifications. Note the small dense image superimposed on the right upper quadrant (arrow).

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 37: Same patient as Fig. 36. Abdominal US (a, b and c) shows echogenic free fluid in the abdominal cavity (a) extending to the scrotum (b and c) where echogenic foci are seen.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Fig. 38: Asymptomatic neonate with intestinal hyperrefringency at prenatal US. Soft abdomen on physical examination. Radiography (a) shows a normal bowel gas pattern. Abdominal US (b) reveals linear peritoneal calcifications (arrow) with no bowel dilatation or free abdominal fluid. The compatible diagnosis in this asymptomatic patient was meconium peritonitis with intrauterine resolution.

© Radiology, Hospital Materno-Infantil Vall d'Hebron - Barcelona/ES
Conclusion

- Radiologists have an important role in the diagnosis of congenital upper gastrointestinal tract abnormalities, although the imaging features may be challenging.
- Awareness of these common and uncommon abnormalities and its associations will guide radiological investigation and lead to an accurate diagnosis, which may be crucial for prompt, adequate treatment of conditions that are life-threatening in some cases.
Personal information

A. Coma MD
Pediatric Radiology Department. Hospital Universitari Materno-Infantil Vall d'Hebrón. Barcelona, Spain.
acoma@vhebron.net

G. Enríquez MD
Institut de Recerca Hospital Vall d'Hebrón. Barcelona, Spain.

I. Barber MD
Pediatric Radiology Department. Hospital Universitari Materno-Infantil Vall d'Hebrón. Barcelona, Spain.

A. Castellote MD
Pediatric Radiology Department. Hospital Universitari Materno-Infantil Vall d'Hebrón. Barcelona, Spain.

J. Piqueras MD
Pediatric Radiology Department. Hospital Universitari Materno-Infantil Vall d'Hebrón. Barcelona, Spain.

E. Vázquez MD
Pediatric Radiology Department. Hospital Universitari Materno-Infantil Vall d'Hebrón. Barcelona, Spain.
References

