Intestinal pathology in neonatal period

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Learning objectives

Findings of bowel disorders are not uncommon in newborns. Consequently, both specialist paediatric radiologists and general radiologists (who may encounter with this sort of patient in an emergency) must be familiar with the imaging features of these diseases, along with the diagnostic techniques used in the assessment.

Bowel disorders present in newborns are specific for this age group. Therefore, diagnostic methods used in these patients will differ from those used in adults or even older children and different imaging findings will be seen.

This presentation will show some examples of the most common bowel diseases in newborns, along with the imaging techniques used for the diagnosis of these disorders.
Background

UPPER INTESTINAL OBSTRUCTION:

Upper intestinal obstruction is the most common abdominal emergency in newborns.

This category will include both gastric obstructions, manifesting as the "single bubble" sign, and duodenal obstructions, leading to the "double bubble" sign.

"Single bubble" sign:

"Single bubble" sign, related to an obstruction in the gastric antral or pyloric region, is an uncommon imaging finding.

Three different kinds of atresia may be identified:

- Atresia with a fibrous band connecting the stomach and duodenum
- Complete atresia with no connection
- Membranous atresia

All of them will exhibit the "single bubble" sign several hours after birth. This sign consists of the finding of a dilated air-filled stomach Fig. 1 on page 9 on abdominal radiograph. Distended air-filled oesophagus may also be present and, in case of incomplete obstruction, some air may be seen distal to the stomach.

Abdominal plain radiograph is enough for diagnosis, contrast-enhanced imaging being contraindicated as it facilitates vomit and bronchoaspiration.

"Double bubble" sign:

The "double bubble" sign, more common than the previous sign, is the manifestation of a duodenal obstruction associated with dilatation of the gastric camera and duodenal bulb.

The etiologic classification is based on the presence or absence of air distal to the obstruction. In absence of air, the cause of the obstruction is duodenal atresia, whereas the presence of distal air will be consistent with a duodenal stenosis, extrinsic compression of the duodenum from annular pancreas, intestinal malrotation with midgut volvulus or peritoneal bands.

DUODENAL ATRESIA:

The duodenum is the most common site of atresia, being atresia far more common than stenosis. The cause is an absence of re-canalization or a partial re-canalization of the duodenum, which is a solid structure during the embryologic development.
About 80% of atresias are located in the proximity of the ampulla of Vater. Consequently, the main diagnostic sign is bilious emesis in the first hours of life and association with abnormalities of the biliary tract is not rare.

The abdominal radiograph, usually diagnostic in these patients, demonstrates the presence of air in the stomach or duodenum ("double bubble" sign) Fig. 2 on page 9, with or without air distal to the obstruction. Performance of other imaging tests is rarely indicated, as the radiologic image is the same either in black (air) or in white (barium).

ANNULAR PANCREAS:

An annular pancreas is due to the persistence of the ventral bud of the pancreas, which encircles the second portion of the duodenum.

An annular pancreas always occurs with a duodenal atresia or stenosis, which may be considered the cause of the obstruction. Therefore, the imaging findings of annular pancreas overlap with those of duodenal atresia or stenosis.

Radiologically, this condition manifests with the "double bubble" sign Fig. 3 on page 10. In those cases where the presence of a concomitant duodenal atresia must be ruled out, a barium study can be performed. This study will allow to determine whether the obstruction is complete or partial Fig. 4 on page 11. Occasionally, the barium column may exhibit and indentation caused by the ring of pancreatic tissue.

DUODENAL BANDS Y ABNORMAL FIXATION OF THE DUODENUM:

Duodenal bands or Ladd's bands are associated with various degrees of intestinal malrotation. They usually produce obstruction to the third or fourth portion of the duodenum. However, obstruction can also occur at higher levels.

The bands usually run from a misplaced cecum, crossing the duodenum, to the liver, posterior peritoneum or abdominal wall and may cause some extent of intestinal obstruction.

INTESTINAL MALROTATION AND MIDGUT VOLVULUS:

The normal small bowel is attached from the duodenojejunal junction, in the left upper quadrant, to the cecum, in the right lower quadrant. In presence of intestinal malrotation, junctions are misplaced, small-bowel mesenteries are abnormality short and volvulation is facilitated.

In abdominal plain radiographs, the midgut volvulus exhibits signs of upper intestinal obstruction Fig. 5 on page 12. Barium studies will show the duodenojejunal junction located to the right of the spine, below the duodenal bulb, and the jejunum located in
the right hemiabdomen. Besides, the duodenum will show a "bird beak" or "corkscrew" appearance and there will be no passage of the barium column. Fig. 6 on page 13.

**SMALL BOWEL OBSTRUCTION:**

Small bowel atresia is more common than stenosis and both are more frequent than duodenal atresia and stenosis.

Nowadays, small bowel obstruction is believed to be caused by an ischemic condition secondary to thromboembolism, stress-related vasospasm, foetal hypoxia or intrauterine volvulus. The common presentation as isolated abnormalities associated with wedge-shaped defects of the mesentery supports the ischemic etiology versus a failure in the intestinal re-canalization.

The abdominal radiographs show dilated small bowel loops associated with presence or absence of air distal to the obstruction depending on the degree of obstruction. The immediately proximal loop to the site of atresia or stenosis usually exhibits a disproportionately dilation, showing a bulbous end. Occasionally, the bulbous end may be filled with fluid leading to mass effect and soft-tissue density.

Obstructions affecting the upper jejunum Fig. 7 on page 14 Fig. 8 on page 15 will exhibit only one or two dilated small bowel loops. When located in mid-jejunum Fig. 9 on page 16 Fig. 10 on page 17, more loops will be involved whereas location in distal ileum Fig. 11 on page 18 will manifest with many dilated loops.

Underused colon is a common finding in barium enema.

**LOWER INTESTINAL OBSTRUCTION:**

**HIRSCHSPRUNG DISEASE:**

Hirschsprung disease is a disorder characterized by the absence of intramural ganglion cells in the distal bowel. Aganglionosis extends distally from the area where neuronal migration has been interrupted to the anus. The aganglionosis causes a lack of relaxation in the aganglionic segment that leads to a functional obstruction.

In most patients, the onset of this disease takes place during infancy. However, a small percentage of patients present with chronic constipation during childhood or adolescence.

The disease is three to four times more frequent in males and, for unknown reasons, it is very uncommon in preterm infants. There is a very rare total colonic type that shows genetic predisposition.

The abdominal plain radiograph only demonstrates signs of lower intestinal obstruction. Consequently, diagnosis must be made using barium enema. The most specific sign in barium enema is a transition from a normal to slightly reduced-calibre bowel (aganglionar)
to a distended bowel (ganglionar) Fig. 12 on page 19 Besides, the rectosigmoid index (ratio obtained from dividing the diameter of the rectum by that of the sigmoid colon) is less than 1 in patients with Hirschsprung disease Fig. 13 on page 20 Barium enema may also show ano-rectal dyskinesia, which consists of irregular contractions in the aganglionic segment. Severe cases may exhibit preobstructive colitis associated to oedema and mucosal ulceration Fig. 14 on page 21

RECTUM ATRESIA AND IMPERFORATED ANUS:

Imperforated anus is a clinic diagnosis and the role of the radiologist is restricted to measure the distance between the rectal stump and the skin surface to indicate the suitability of an immediate anus reconstruction or a differed reconstruction with temporary colostomy. The radiologist must also rule out genitourinary malformations, frequently associated to this condition.

Measure of the distance between the rectal stump and the skin surface may be performed with abdominal or perineal ultrasound or with an invertogram procedure consisting of hanging the patient upside-down to allow intestinal air to fill the colonic stump Fig. 15 on page 22.

FUNCTIONAL IMMATURITY OF THE COLON AND MECONIUM PLUG:

This condition is a common cause of lower intestinal obstruction in the newborn and it is believed to be related to a motility dysfunction. The term meconium plug is inadequate as this is not the real cause of obstruction.

The newborns affected by the disease are usually born from a diabetic mother or a mother who has been administered magnesium sulphate as a treatment for eclampsia.

The abdominal plain radiograph shows signs of lower intestinal obstruction Fig. 16 on page 23 The barium enema, which allows the diagnosis, exhibits dilatation of the ascending and transverse colon, a transition close to the splenic flexure and very-small-calibre descending colon, rectum and sigma. Rectum is usually quite distensible and, consequently, the rectosigmoid index should be equal to or greater than 1. Repletion failures of the colon as a result of meconium impaction may be seen. Fig. 17 on page 24

The disease usually shows a benign course and a significant clinical improvement is common after barium enema.

MECONIUM ILEUS:

Meconium ileus refers to a lower intestinal obstruction secondary to abnormal meconium impaction in the colon and distal ileum. This condition is predominantly seen in children with cystic fibrosis.
Abdominal plain radiograph shows signs of lower intestinal obstruction and images of "bubbles" or "bread crumbs" in the right iliac fossa, as a result of the combination of air and meconium. Besides, there is a wide variation in the calibre of the dilated loops and scarce air-fluid levels.

Barium enema demonstrates a microcolon, the smallest calibre being seen in the area of meconium ileus. The 10-30 distal centimetres of ileum present a relatively small, although larger than the colonic, calibre and show round or ovoid repletion failures that correspond to impacted meconium Fig. 18 on page 25.

**NECROTIZING ENTEROCOLITIS:**

This idiopathic enterocolitis is commonly observed in preterm infants. Children who have not received any food rarely present with this condition, so it is possible that milk or formulas facilitate the proliferation of microorganisms. It is also seen in full-time newborns when epidemic breakouts take place, which supports the idea of an infectious etiology.

Abdominal radiographs may show a diffuse dilation of air-filled intestine with asymmetric gas distribution throughout the intestine and alternative areas of distended air-filled loops and loops with slight air content.

Pneumatosis intestinalis and presence of gas in the portal system of a preterm infant are pathognomonic signs of necrotizing enterocolitis Fig. 19 on page 26.

The barium enema will show an irregular and oedematous mucosa.

**MECONIUM PERITONITIS:**

Meconium peritonitis is the result of an intra-uterine bowel perforation, secondary to a large or small bowel obstruction, and the subsequent spillage of foetal meconium into the peritoneal cavity. The irritative meconium triggers an inflammatory reaction that leads to the formation of a fibrous capsule composed of granulation tissue.

Radiologically, this condition manifests at birth as large intra-abdominal cysts with a heterogeneous content and a thick, sometimes calcified, wall Fig. 20 on page 27.

**INTESTINAL PERFORATION IN THE NEWBORN:**

This condition is secondary to an intestinal obstruction or a necrotizing enterocolitis.

The characteristic imaging finding is pneumoperitoneum. This sign may appear in the newborn, when the pneumoperitoneum is massive, as a diffuse hyperclarity in the abdomen ("balloon sign") Fig. 21 on page 28. Gas desiccating the falciform ligament or the medial umbilical ligament and the double wall sign may be identified. Fig. 22 on page 29.
Fig. 1: "Single bubble" sign. Dilated air-filled gastric camera with no air distal to the obstruction, in a patient with gastric atresia. References: Servicio de Radiodiagnóstico, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 2: "Double bubble" sign. Distended gastric camera and duodenal bulb, associated with little air distal to the obstruction in a patient with duodenal stenosis. References: Servicio de Radiodiagnóstico, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 3: "Double bubble" sign in a patient with annular pancreas associated with duodenal atresia. References: Servicio de Radiodiagnóstico, Hospital Universitario Central de Asturias - Oviedo/ES

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**Fig. 4:** Barium study, in the same patient from figure 3, which shows a complete obstruction to the duodenal bulb consistent with the presence of duodenal atresia secondary to annular pancreas. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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**Fig. 5:** Patient with intestinal malrotation and midgut volvulus. Dilated air-filled gastric camera and little air distal to the obstruction. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 6: Barium study. Duodenojejunal junction located to the right of the spine showing with a "corkscrew" morphology, in the same patient from fig. 5. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 7: Fig. 7: Upper jejunal stenosis. Large dilatation of the first jejunal loops with air-fluid levels on the standing radiograph and presence of little air and faeces distal to the obstruction. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 8: Upper jejunal stenosis. Dilatation of the first jejunal loops with air distal to the obstruction on the abdominal plain radiograph. Barium enema shows an underused colon. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 9: Fig. 9: Mid-jejunal stenosis. Large dilatation of several small bowel loops with air-fluid levels on the standing radiograph. Presence of air distal to the obstruction confirms a partial obstruction. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 10: Barium enema in the same patient from fig. 9. Underused colon.
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**Fig. 11:** Fig. 11: Ileum atresia. Multiple dilated small bowel loops with air-fluid levels on the standing radiograph. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 12: Fig. 12: Patient with Hirschsprung disease. Dilated colon with presence of abundant faeces on the abdominal plain radiograph. Barium enema demonstrates a change in the calibre of the sigmoid colon, showing a normal-calibre distal segment and a very dilated proximal segment. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 13: Fig. 13: Barium enema in the same patient from fig. 12. Lateral projection that shows a rectosigmoid index of less than 1. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 14: Severe Hirschsprung disease with preobstructive colitis. Severe oedema of the colonic mucosa. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 15: Imperforated anus. Abdominal plain radiograph that exhibits dilatation of small bowel loops and colon to the rectal ampulla. Invertogram to measure the distance from the rectal stump to the skin. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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**Fig. 16:** Functional immaturity of the colon. Dilated small bowel loops and ascending colon. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 17: Fig. 17: Barium enema in the same patient from figure 16 showing transition in the transverse colon, close to the splenic flexure. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 18: Fig. 18: Patient with meconium ileus. The abdominal radiograph exhibits dilated small bowel loops in absence of air distal to the obstruction and barium enema shows a small-calibre colon with multiple repletion defects secondary to meconium impaction.

References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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**Fig. 19:** Fig. 19: Patient with diffuse pneumatosis and necrotizing enterocolitis. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 20: Patient with colon atresia and intra-uterine perforation associated with a large meconium cyst. The abdominal radiograph shows a significant mass effect with soft tissue density involving almost the whole abdomen and peripheral curvilinear calcifications. The ultrasound image demonstrates the presence of a large thick-walled cyst of inhomogeneous content with calcifications. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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Fig. 21: Fig. 21: "Balloon sign" in a patient with perforated necrotizing enterocolitis and massive pneumoperitoneum. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES

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**Fig. 22:** 'Fig. 22: Lateral decubitus radiograph in a patient with intestinal perforation secondary to enterocolitis. "Double wall" sign. References: Radiology Department, Hospital Universitario Central de Asturias - Oviedo/ES'

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Abdominal plain radiograph is enough for diagnosis in upper intestinal obstruction.

In lower intestinal obstruction as in Hirschsprung disease, functional immaturity of the colon or meconium ileus, barium enema may be useful.
Conclusion

Bowel disorders present in newborns are specific for this age group. Therefore, clinical and radiologic manifestations in newborns will differ from those seen in adults and older children with bowel disorders, and different diagnostic methods will be used.

Imaging studies commonly allow the diagnosis in these patients and point to the appropriate treatment. Consequently, knowledge of these studies is vital for both specialist paediatric radiologists and general radiologists.
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