Intestinal obstruction and the double bubble sign

Intestinal obstruction in a newborn infant is associated with numerous aetiologies. Following a careful clinical examination, gastrointestinal imaging can reveal the presence of the ‘double bubble’ sign on an abdominal X-ray. Various conditions can result in a double bubble; being aware of these conditions and recognising their imaging pattern will help in early diagnosis and timely and appropriate management.

**Key points**

- Infants presenting with vomiting, aspiration, abdominal distension and/or feeding intolerance in the neonatal period should be investigated for intestinal obstruction.
- The classic ‘double bubble’ sign is seen on an abdominal X-ray in a number of obstructive anomalies.
- The help of a radiologist will be necessary. Identifying these conditions promptly will ensure timely diagnosis and management.

**Keywords**
gastrointestinal imaging; double bubble; neonate; duodenal obstruction

---

The double bubble sign in newborn infants is seen on plain abdominal radiographs in a variety of conditions. It depicts two air-filled structures in the upper abdomen and represents a gas-distended dilated stomach and proximal duodenum, implying congenital proximal bowel obstruction.

The specific anomaly causing the obstruction will determine the appearance and presence of air distal to the obstruction. A neonate with a double bubble and no distal bowel gas (complete obstruction) can be presumed to have duodenal atresia and in most cases no further imaging is required. However, if double bubble is present with distal bowel gas (partial obstruction) the differential diagnosis includes duodenal stenosis, duodenal web, annular pancreas and midgut volvulus. In these cases usually an upper gastrointestinal (GI) contrast study is performed to distinguish between midgut volvulus, which requires emergent surgery, and other causes that can be repaired electively.

**Duodenal atresia**

The most common upper intestinal obstruction in neonates is duodenal atresia, in which the duodenum does not develop properly during embryogenesis resulting in a closed off tube that does not allow the passage of the stomach contents (FIGURE 1). It is associated with additional anomalies in >50% of cases, for example VACTERL association (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities), shunt vascularity cardiac lesions (atrial septal defect, ventricular septal defect, patent ductus arteriosus) and annular pancreas. Thirty per cent of babies with duodenal atresia have Down's syndrome.

Infants with duodenal atresia present early in the neonatal period with abdominal distension, vomiting and absent bowel movements. If atresia is distal to the ampulla of Vater the vomiting is bilious.

---

**FIGURE 1** (A) Duodenal atresia and the double bubble. (B) A plain X-ray of a two-day-old infant born at 36 weeks' gestation showing the double bubble appearance in the upper abdomen with no gas seen distally. The baby had respiratory distress, sepsis and jaundice.
If the atresia is proximal to the ampulla, the vomiting is non-bilious. With surgical treatment the survival rate is >90%.

**GI imaging**
- Plain radiographs reveal the double bubble sign without distal bowel gas (FIGURE 1). Rarely, a small amount of gas may be seen in the distal GI tract in the presence of biliary/pancreatic duct variations allowing bowel gas to travel through the ampulla of Vater and enter the distal bowel. If the stomach or duodenum is decompressed by a nasogastric tube or vomiting, the double bubble may not be seen on initial radiographs.
- Ultrasound reveals a dilated fluid- and gas-filled stomach and duodenum.
- An upper GI contrast study shows contrast within the distended stomach and proximal duodenum with no contrast seen distally, however, the study is not usually performed because radiographs are essentially diagnostic.

**Duodenal stenosis**
Duodenal stenosis is a less severe variant of a duodenal anomaly in which there is partial recanalisation of the duodenum during embryogenesis and fixed narrowing of the duodenal lumen. As with duodenal atresia, duodenal stenosis is associated with additional anomalies in >50% of cases.

Duodenal stenosis results in a variable and often delayed clinical presentation due to the incomplete nature of the obstruction. Infants may present with recurrent episodes of vomiting, aspiration or failure to thrive.

**GI imaging**
- Plain radiographs reveal the double bubble sign with distal bowel gas (FIGURE 2).
- Ultrasound reveals a dilated fluid- and gas-filled stomach and duodenum.
- An upper GI contrast study shows a focal or longer segment of fixed circumferential narrowing (FIGURE 2).

**Duodenal web**
A duodenal web refers to a membranous web or intraluminal diverticulum resulting in complete or incomplete obstruction at the duodenum. It is associated with Down's syndrome, malrotation and annular pancreas. It presents clinically as feeding intolerance, vomiting (bilious more than non-bilious), weight loss and dehydration. There is an excellent prognosis with treatment.

**GI imaging**
- Plain radiographs reveal the double bubble sign with distal bowel gas (FIGURE 3).
- Ultrasound reveals a dilated fluid- and gas-filled stomach and duodenum. Fluid may outline a thin web or show a hypoechoic intraluminal diverticulum.
- An upper GI contrast study typically gives the windsock sign (FIGURE 3B), which represents an intra-duodenal barium contrast-filled sac surrounded by a narrow lucent line (web or intraluminal mucosal diverticulum).

**Annular pancreas**
An annular pancreas is a developmental anomaly in which a ring of pancreatic tissue surrounds the second portion of the
duodenum. It can be complete if the pancreatic parenchyma surrounds the duodenum or incomplete when the duodenum is not completely surrounded. It is associated with additional anomalies such as Down’s syndrome, pancreas divisum, duodenal stenosis and malrotation. Newborn infants present with signs of duodenal obstruction with typically non-bilious vomiting, feeding intolerance and abdominal distension.

**GI imaging**
- Plain radiographs reveal the double bubble sign with distal bowel gas.
- Ultrasound reveals a dilated fluid-filled duodenum passing through the pancreas (FIGURE 4).
- An upper GI contrast study gives a dilated stomach and first portion of the duodenum, with delayed emptying (FIGURE 4). Circumferential narrowing of the second portion of the duodenum is seen.
- Magnetic resonance cholangiopancreatography (MRCP: a scan that specifically images the pancreatic and bile ducts) shows the pancreatic duct encircling the duodenum.

**Malrotation and midgut volvulus**
Malrotation is an abnormality of the bowel – a failure of the normal rotation of the bowel during embryogenesis. Midgut volvulus is a complication of a malrotated bowel in which the intestine twists so that the blood supply to that part of the bowel is cut off; it is a surgical emergency with a high mortality rate and a delay in diagnosis can result in diffuse bowel necrosis and death. The newborn infant often appears entirely normal for a period of time before suddenly presenting with bilious vomiting.

**GI imaging**
- Plain radiographs may be normal but most commonly reveal multiple dilated loops of bowel. Less commonly they may show a double bubble, however, the dilated proximal duodenum is not markedly enlarged as usually seen in a longstanding obstruction such as duodenal atresia.
- Ultrasound reveals a clockwise whirlpool sign, seen when a structure twists on itself. In this instance, the bowel rotates around its mesentery with the mesenteric vessels creating the whirls (best seen on Doppler). An inverted superior mesenteric artery (SMA)-superior mesenteric artery (SMA) sentinel vessel.
An upper GI contrast study is the investigation of choice if malrotation/midgut volvulus is suspected. The aim of the study is to exclude or demonstrate findings of malrotation (with or without volvulus) in newborn infants with bilious vomiting. On anteroposterior projection, the duodenojejunal flexure is seen to the right of the left sided vertebral body pedicle and inferior to the duodenal bulb. On lateral projection, the second-third portion of the duodenum is not located posteriorly in the retroperitoneal position. In the presence of volvulus there is a spiral appearance of the distal duodenum and proximal jejunum – the corkscrew sign – and this is diagnostic for midgut volvulus.

**Conclusion**

Various conditions can result in a double bubble sign and management differs from one condition to another. Recognising these conditions by their imaging pattern will ensure an early diagnosis and subsequent timely management.

**Further reading**