The double bubble sign

Dr B A Sethi
Specialist registrar radiology
Dr K A Duncan
Consultant radiologist
Mr C P Driver
Consultant paediatric surgeon
Royal Aberdeen Children’s Hospital, Aberdeen, UK

Introduction
The double bubble sign in neonates is seen on plain abdominal radiographs in a variety of conditions. It represents gas distended dilated stomach and proximal duodenum, and implies congenital proximal bowel obstruction. A newborn with a double bubble and no distal bowel gas can be presumed to have duodenal atresia and in most cases no further imaging is required. If double bubble is present with distal bowel gas the differential diagnosis includes duodenal stenosis, duodenal web, annular pancreas and midgut volvulus. In these cases usually an upper GI contrast study is performed to distinguish between midgut volvulus which requires emergent surgery, and other causes which can be repaired electively.

Duodenal atresia
• The most common upper intestinal obstruction in neonates and represents the lack of recanalisation of the duodenum during embryogenesis.
• Associated with additional anomalies in >50% of cases – VACTERL, shunt vascularity cardiac lesions (atrial septal defect, ventricular septal defect, patent ductus arteriosus), annular pancreas; 30% of babies with duodenal atresia have Down’s syndromes.
• With surgical treatment survival rate is >90%.
• Clinical presentation: Present early in life with abdominal distension, vomiting and absent bowel movements. If atresia is distal to the ampulla of Vater vomiting is bilious. If atretic segment is proximal vomiting is non-bilious.
• Plain radiographs: Double bubble without distal bowel gas. Very 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 NG tube or vomiting, the double bubble may not be seen on initial radiographs.
• Ultrasound: Dilated fluid and gas filled stomach and duodenum.
• Upper GI contrast study: Not usually performed because radiographs are essentially diagnostic.

Duodenal stenosis
• A less severe variant of a duodenal anomaly representing partial recanalisation of the duodenum with fixed narrowing of the duodenal lumen.
• As with duodenal atresia, duodenal stenosis is associated with additional anomalies in >50% of cases.
• Clinical presentation: Results in variable and often delayed presentation due to the incomplete nature of obstruction. Children may present with recurrent episodes of vomiting, aspiration or failure to thrive.
• Plain radiographs: Double bubble with distal bowel gas.
• Ultrasound: Dilated fluid and gas filled stomach and duodenum.
• Upper GI contrast study: Focal or longer segment of fixed circumferential narrowing.

Duodenal web
• Refers to a membranous web or intraluminal diverticulum resulting in complete or incomplete obstruction at the duodenum.
• Associated with Down’s, malrotation and annular pancreas.
• Excellent prognosis with treatment.
• Clinical presentation: Incomplete obstruction resulting in vomiting (bilious > non-bilious), weight loss and dehydration.
• Plain radiographs: Double bubble with distal bowel gas.
• Ultrasound: Dilated fluid and gas filled stomach and duodenum. Fluid may outline a thin web or show a hypoechoic intraluminal diverticulum.
• Upper GI contrast study: Typically gives the whirlpool sign which represents an intra-duodenal barium contrast filled sac surrounded by a narrow lucent line (web or intraluminal mucosal diverticulum).

Annular pancreas
• Represents a developmental anomaly where a ring of pancreatic tissue surrounds the second portion of duodenum. It can be complete where the pancreatic parenchyma completely surrounds the duodenum or incomplete where the duodenum is not completely surrounded.
• Associated with additional anomalies such as Down’s, pancreas divisum, duodenal stenosis and malrotation.
• Clinical presentation: Newborns present with signs of duodenal obstruction with typically non-bilious vomiting, feeding intolerance and abdominal distension.
• Plain radiographs: Double bubble with distal bowel gas.
• Ultrasound: Dilated fluid filled duodenum passing through the pancreas.
• Upper GI contrast study: Dilated stomach and first portion of duodenum, with delayed emptying. Circumferential narrowing of second portion of duodenum.
• MRCP: Shows the pancreatic duct encircling the duodenum.

Malrotation and midgut volvulus
• Malrotation is failure of normal rotation of bowel during embryogenesis, predisposing to volvulus due to abnormal mesenteric fixation. Volvulus is a life threatening complication of malrotated bowel.
• It is a true surgical emergency with a high mortality rate. Delay in diagnosis can result in diffuse bowel necrosis and death.
• Clinical presentation: Neonate is often entirely well for a period of time before suddenly presenting with green bile vomiting.
• Plain radiographs: May be normal. Most commonly appears as multiple dilated loops of bowel. Less commonly may produce a double bubble. The dilated proximal duodenum is not markedly enlarged as usually seen in longstanding obstruction such as duodenal atresia.
• Ultrasound: Clockwise whirlpool sign seen when structures twist on themselves. In this instance bowel rotates...
around its mesentery with the mesenteric vessels creating the whirls, best seen on Doppler. Inverted SMA-SMV relationship with the SMV to the left of the SMA may be seen.

- Upper GI contrast study: 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 neonates with bilious vomiting. Typically on the AP projection the DJ flexure is seen to the right of the left sided vertebral body pedicle and inferior to the duodenal bulb. On lateral projection, the second to third portion of 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, this is diagnostic of midgut volvulus.

**Conclusion**

Various conditions can result in a double bubble sign and management differs from one condition to another. Being aware of these conditions and recognising their imaging pattern will help in early diagnosis and subsequent timely management.

**Further reading**

8. https://my.statdx.com/document/annular-pancreas/93bd8984-5161-45e4- a8c4-a5257b6a7d5/searchTerm=double bubble sign
9. https://my.statdx.com/document/malrotation/36d04e12-b01c-48a1-9129- fa5f873990f/searchTerm=double bubble sign

**Figure 1**

Duodenal atresia. Two-day-old born at 36 weeks. On delivery RDS, sepsis, jaundiced. (A) Plain radiograph: Double bubble appearance in the upper abdomen with no gas seen distally. (B) Upper GI contrast: Contrast lies within the distended stomach and proximal duodenum with no contrast seen distally.

**Figure 2**

Duodenal stenosis. One-day-old term baby. Bilious vomiting, suspected sepsis. (A) Plain radiograph: Double bubble appearance with gas seen in the jejunal loops. (B) Upper GI contrast study: Contrast opacification of the stomach and dilated D1 is seen. There is circumferential narrowing of the D2. Distal duodenum crosses the midline.
Figure 3
Duodenal web. Term baby. AVSD and double bubble on antenatal scans. Morphological features of trisomy 21. (A) Plain radiograph: Moderately dilated stomach and duodenum with gas seen in the distal loops. (B) Upper GI contrast study: Ballooning and dilatation of the proximal duodenum with only a very small trickle of contrast identified to pass into the distal duodenum after approximately 40 minutes (C).

Figure 4
Annular pancreas. Eight-day-old baby with persistent non bilious vomiting since birth. Dehydrated with metabolic alkalosis. (A) Ultrasound abdomen: Performed to rule out pyloric stenosis. Normal pylorus. Distended fluid filled viscus in the region of the pylorus/duodenum, raising the suspicion of duodenal obstruction. (B and C) Upper GI contrast study: On initial screening a double bubble appearance with dilatation of the first part of the duodenum, with tiny amount of gas in the distal bowel. On injecting contrast, the contrast outlines the stomach and dilated first and second part of the duodenum with no drainage distally.
Malrotation and midgut volvulus. One-day-old baby born at 36 weeks. Bilious vomiting, very poor feeding.
(A) Plain radiograph pre-malrotation, non specific bowel gas pattern with gas seen within the distal bowel.
(B) Plain radiograph post malrotation: Double bubble sign with no gas distally. (C) Upper GI contrast study: Contrast confirmed in small bowel in the right side of the abdomen with no evidence of crossing the midline to the left. Appearances are in keeping with malrotation.