Anal atresia, i.e. imperforate anus, is not an infrequently seen congenital anomaly with an average worldwide incidence of 1:5,000 live births. Congenital jejunoileal obstruction caused by atresia or stenosis is also a relatively common cause of neonatal intestinal obstruction, with an incidence of 1:400 to 1:1,500 live births. In contrast, colonic atresia is a rare cause of intestinal obstruction in neonates, with an incidence of about 1:20,000 live births. Most neonates with imperforate anus have one or more abnormalities that affect other systems. However, only two of 172 patients with imperforate anus were reported to have an intestinal atresia below the duodenum, and only two of 246 patients had an absent colon. It is thus extremely rare to see a patient with an imperforate anus and concurrent ileal stenosis and colonic atresia, as described by Asabe and Nagasaki in this issue of the Asian Journal of Surgery.

The aetiology of intestinal or colonic atresia is not clearly understood. The current consensus is that there is an in utero mesenteric vascular catastrophe, such as volvulus, intussusception, incarceration, or strangulation secondary to hernias, and embolic or thrombotic events, which results in ischaemia and absorption of the necrotic bowel segment. The cause of imperforate anus is regarded as an event in the abnormal embryological development that results from defects in the shape of the dorsal (posterior) cloaca and an absence of the dorsal cloacal membrane. The lack of relationship between the causes of intestinal atresia and imperforate anus may, thus, explain the rarity of their coexistence.

The level of rectal atresia in imperforate anus has traditionally been assessed by an invertogram, as in the reported case. In our institute, we rarely use the invertogram for the diagnosis of congenital anorectal malformation. Instead, magnetic resonance imaging (MRI) of the pelvis is regularly performed for direct visualization of the distal rectal pouch. On the invertogram, the presence of an obstruction proximal to the rectum, such as stenosis or atresia of the small bowel or colon, will obviously result in a "high-held" terminal gas shadow because the intraluminal gas would pause proximally without reaching the very terminal part of the rectum. However, there are still no reports in the literature regarding MRI findings in anorectal malformation with concurrent intestinal atresia. Moreover, lower gastrointestinal (GI) contrast study is certainly not applicable in patients with imperforate anus, and the absence of an antenatal history of polyhydramnios does not exclude the presence of intestinal atresia either. Therefore, correct preoperative diagnosis in such patients is difficult to make and inadequate initial management often occurs as a consequence, as reported by Asabe and Handa.

We recently encountered a patient with concurrent oesophageal atresia and imperforate anus, in whom preoperative MRI clearly demonstrated the level of atresia in both segments. Division of the tracheo-oesophageal fistula with primary oesophageal anastomosis and creation of colostoma for the rectal atresia were successfully performed in the same setting. However, emergency laparotomy was conducted the next day because postoperative abdominal X-ray films showed persistence of a bowel gas shadow with a progressively increasing size at the right upper quadrant. Surgical findings confirmed that there was volvulus of the terminal ileum due to torsion of Meckel's diverticulum along the axis of the omphalomesenteric duct. In this case, volvulus of the ileum possibly occurred only after GI continuity was re-established by oesophageal anastomosis which allowed air entry into the distal bowel.
In summary, lower intestinal atresia associated with imperforate anus is difficult to diagnose preoperatively and often necessitates a second operation. It is therefore important, as suggested by Asabe and Nagasaki,6 that a vigorous search for the presence of another GI obstruction should be performed at the first laparotomy when the terminal gas shadow on an invertogram is too high up in this group of patients.

References