Case report	
	
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Title	A RARE COEXISTENCE OF INTESTINAL ATRESIA AND BILIARY ATRESIA
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Keywords	**Abstract**
Biliary Atresia, Intestinal Atresia, Malrotation, Meconium Peritonitis	Intestinal atresia is one of the rare congenital anomalies in which there is a total disruption in the continuity of gastrointestinal tract. It is often associated with other conditions but its association with biliary atresia is not well known. We present two patients who were initially diagnosed with intestinal atresia and associated gastrointestinal anomalies (case 1 had type III duodenal

Abbreviatios Duodenal Atresia (DA), Total Parenteral Nutrition (TPN), Intestinal Failure Associated Liver Disease (IFALD), Biliary Atresia (BA), Percutaneous Cholangiogram (PTC), Hepatobiliary iminodiacetic acid (HIDA)	atresia with annular pancreas, malrotation variant without volvulus, and ascending colon atresia; while case 2 had meconium peritonitis secondary to intrauterine volvulus, and distal jejunal atresia type I), both underwent respective corrective surgeries, and later on found out to have biliary atresia as well. None of these patients proceed with Kasai portoenterostomy taking into consideration that the patients had previous laparotomy for intestinal atresia. Relaparotomy poses significant challenges due to prior bowel resection and the risk of limited remaining bowel length required to maintain adequate growth and development. The coexistence of biliary atresia further complicates management, as Kasai portoenterostomy in this context is technically challenging and may require modification. There are few reported cases of intestinal atresia with biliary atresia in which Kasai portoenterostomy was performed, but those patients eventually succumbed to death. Concurrent intestinal atresia with biliary atresia although uncommon, is important to take note of the possibility of these associations.

Introduction

Intestinal atresia is one of the rare congenital anomalies in which there is a total disruption in the continuity of gastrointestinal tract. It is one of the commonest causes of neonatal intestinal obstruction, occurs in approximately 1 per 5,000 live births[(1)]. By far the most widely accepted description on the pathophysiology of duodenal atresia is by Viennese anatomist, Dr Julius Tandler who hypothesised the failure of recanalization of solid-cord duodenum during embryogenesis, whereas for jejunoileal atresia, the hypothesis is the intrauterine ischemic insult to the midgut, affecting single or multiple segments of the already developed intestine[1]. Intestinal atresia is often associated with other abnormalities but its association with biliary atresia is not well known.

Case report

Case 1: Baby X, a term male neonate was referred to us for antenatally detected double-bubble sign and polyhydramnios. Amniocentesis revealed a normal 46XY. The patient was born term at 38 weeks 1 day via elective lower segment caesarian section, with good APGAR score and birth weight of 2150g, to a 39-year-old Para 3 mother who had gestational diabetes mellitus on diet control and pregnancy-induced hypertension. Clinically the child was not dysmorphic, had clear orogastric aspirate, normal per abdominal and genital examinations, and a patent anus. Plain abdominal radiograph with air insufflation (figure 1) showed double bubbles without distal bowel gas. These findings are consistent with duodenal atresia (DA). The operation was delayed to day 15 of life as the child was tested COVID-19 positive and required quarantine.

Laparotomy was performed, and intra-operatively we found out that the child has type III DA with annular pancreas, malrotation variant without volvulus, and ascending colon atresia (figure 2A). Division of Ladd's band, broadening of small intestinal mesentery, and

side-to-side duodenoduodenostomy were performed. Ileum and caecum appeared to be congested after releasing dense adhesion at the root of the mesentery, in which part of the ileum showed sign of viability after applying warm pack (figure 2B). The atretic part of ascending colon was resected, together with non-viable caecum, appendix, and terminal ileum; proximal stump was clipped and dropped, distal stump was brought out as distal stoma, and laparostomy was created. Second-look operation was performed 48 hours later; a total of 25cm small bowels improved after resuscitation, 45cm small bowels remained gangrenous (figure 2C) and hence resected, and distal jejunum was brought out as proximal loop of stoma (figure2D). Total viable bowels left is 75cm from the site of duodenoduodenostomy. Reversal of stoma was performed one month after the second-look operation.

Feeding was initiated 2 weeks after the second-look operation, but the child had feeding intolerance and required a long duration of total parenteral nutrition (TPN), which he later developed into type 2 intestinal failure associated liver disease (IFALD). TPN was withheld for a few days, however serum bilirubin level remains elevated (total bilirubin level ranging 80-180umol/L, direct bilirubin level ranging 60-130umol/L) and transaminitis did not improve. Also, we noticed that the child never had pigmented stool since initiation of feeding.

Hepatobiliary ultrasound at day 94 of life showed well-distended gallbladder without gallbladder wall thickening, presence of triangular cord sign, thickened hepatic artery measuring 0.2cm, and multiple hypoechoic liver lesions scattered in right liver lobes, which raised the suspicion of biliary atresia (BA), hence proceeded with percutaneous cholangiogram (PTC) and liver biopsy. PTC showed neither flow of contrast into duodenum nor intrahepatic ducts (figure 3); liver biopsy histopathological examination showed lymphocytic and neutrophilic infiltration of portal tracts, associated with portal fibrosis, edema and prominent ductular reaction, bile plugs are seen within proliferating bile ducts and bile ductules, with evidence of bridging fibrosis (figure 4); all these findings are significant features of BA.

Family conference was held, and parents agreed with conservative management in view of the poor prognosis and Kasai portoenterostomy may not be worth it, taking into consideration that the patient had undergone laparotomy for intestinal atresia, the challenges of re-laparotomy secondary to severe adhesion from previous laparotomy and he relatively short bowel length remains post bowel resection and anastomosis. The patient had worsening liver failure and succumbed to death at 6 months old.

Case 2: Baby Y, a female newborn was referred to us for antenatally detected polyhydramnios and suspected bowel perforation. The patient was born prematurely at 31 weeks 3 days via emergency lower section caesarean section for dichorionic diamniotic twin pregnancy in labour, with a birth weight of 1.75kg, to a 27-years-old Para 2 mother who had gestational diabetic mellitus on diet control. She was born not vigorous with poor APGAR score and was intubated shortly after birth due to poor respiratory effort. Clinically the patient was not syndromic, her abdomen was distended and had bilious orogastric aspirate. Genital and perineal examinations were normal. Plain abdominal radiograph (figure 5) showed presence of stomach and proximal bowel gases and absence of distal bowel gas, with calcifications seen at both upper quadrants of the abdomen.

Laparotomy was performed on day 2 of life. Upon entering into the peritoneum, pus and meconium came into view; there was severe interloop slough and bowel adhesions, and calcification was seen all over the abdomen sparing the left lower quadrant of the abdomen. Segmental jejunal volvulus was detected 33cm from duodeno-jejunal junction and 55cm from the ileocaecal valve, in which the involved bowel was gangrenous and perforated (Figure 6). Distal jejunum was atretic. 30cm of gangrenous bowel was resected and both ends of the small bowel loops were brought out as jejunostomy.

Post-operatively the patient was unable to achieve full enteral feeding and developed short bowel syndrome. The high stoma output, coupled with her underlying large patent ductus arteriosus in failure, resulted in poor weight gain. She was put on prolonged TPN for nutritional rehabilitation, before undergoing reversal of stoma 2 months after, and full enteral feeding was established 2 weeks thereafter. During that period of time, the child developed TPN cholestasis and worsening transaminitis. Refeeding was initiated to stimulate enterohepatic circulation and distal gut formation, and urso-deoxycholatic acid was given to promote bile secretion and bile flow from the liver, but despite that, the child's serum bilirubin level remains elevated (total bilirubin level ranging 100-140umol/L, with direct bilirubin level

ranging 60-100umol/L) and she started passing acholic stool after stoma reversal. Clinically the liver is slightly enlarged and firm in consistency.

Hepatobiliary ultrasound at day 68 and day 82 of corrected gestational age showed some features that may be suggestive of biliary atresia. The patient underwent Hepatobiliary iminodiacetic acid (HIDA) scan later on, which showed good tracer uptake by the liver but the gallbladder was unable to be visualised and there was no tracer activity demonstrable in the gastrointestinal tract throughout the HIDA study. Liver biopsy and cholangiogram were not performed as the parents were not keen on the procedures. The child later develops decompensated liver failure, portal hypertension and oesophageal varices and managed symptomatically.

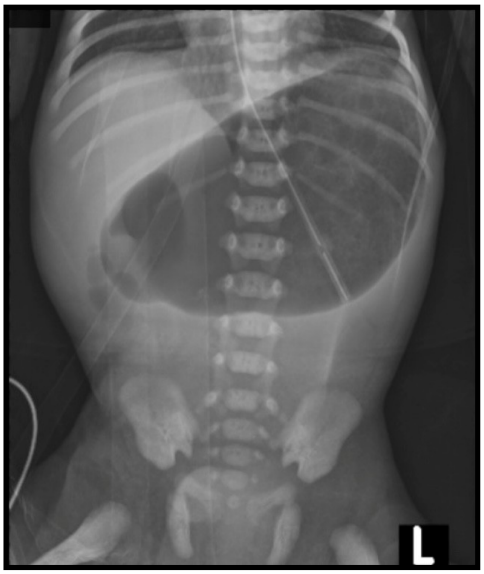

Figure 1 | Plain abdominal radiograph with 20ml air insufflation taken on day 7 of life showed double-bubble sign without distal bowel gas

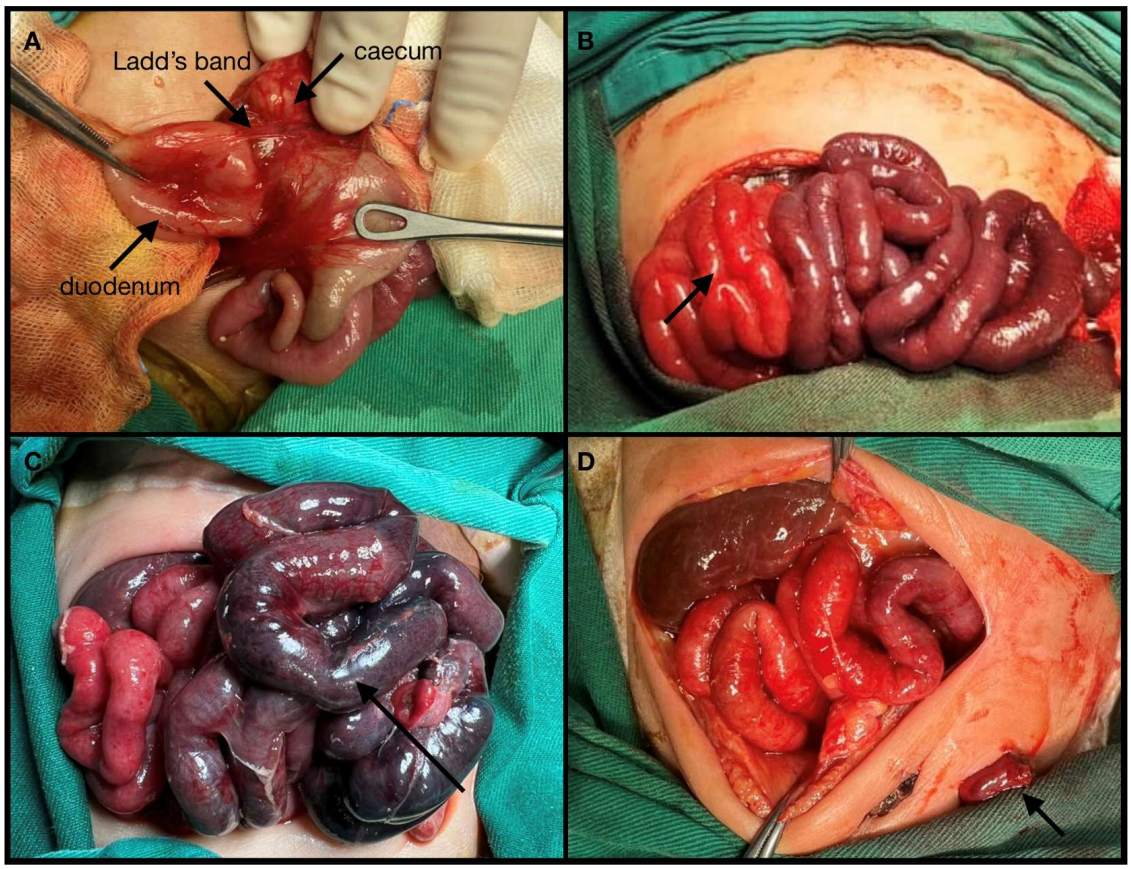

Figure 2 | (A) Intraoperative photograph taken during first laparotomy: caecum was found right next to the left of duodenum, with Ladd's band connecting the two structures. **(B)** Ileum appeared congested after releasing dense adhesion at the root of mesentery, in which part of the distal ileum (arrow) showed signs of viability after applying warm pack, hence decision of laparostomy was made for bowel resuscitation. **(C)** 45cm of ileum was found to be remained gangrenous (arrow) during second-look operation, and was resected. **(D)** Total of 75cm viable small bowel was left after second-look operation; distal jejunum was brought out as proximal loop of stoma at left iliac fossa region, medial to the distal loop created during first laparotomy (arrow).

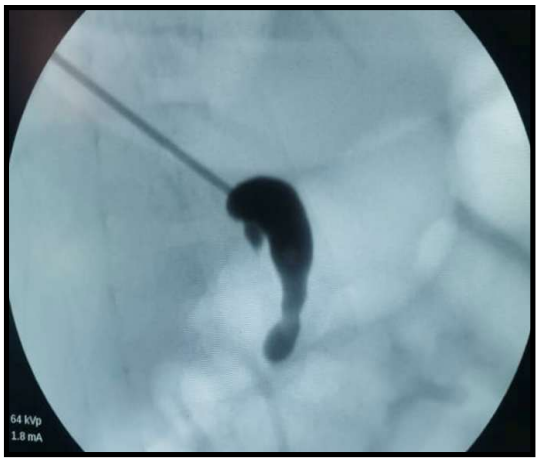

Figure 3 | Percutaneous cholangiogram was performed on day 100 of life. 18G co-axial needle was introduced into the gallbladder under aseptic technique with ultrasound guidance, until bile reflux was seen.15ml of Omnipaque contrast was injected, opacification of contrast seen filling up the lobulated gallbladder. No opacification of cystic, common hepatic, and common bile ducts seen

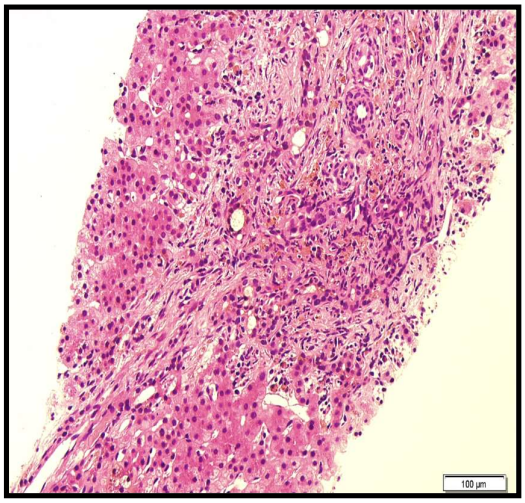

Figure 4 | Liver biopsy (with 40x magnification) shows portal tracts with a moderate infiltration by lymphocytes and neutrophils, associated with fibrosis, oedema and prominent ductular reaction. The hepatocytes reveal mild to moderate hepatocanalicular cholestasis.

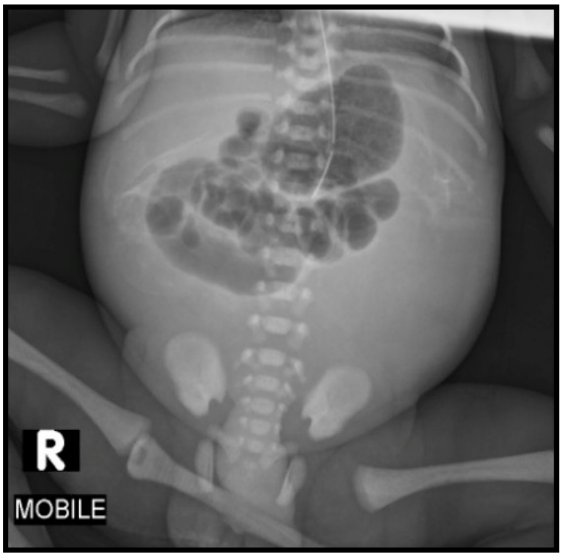

Figure 5 | Plain abdominal radiograph taken at 17 hours of life showed stomach shadow with proximal bowel gas and absence of distal bowel gas. Ryle's tube was placed into the stomach. Calcifications were seen at both upper quadrants. Radio-opaque object near the genital region is the umbilical cord clamp.

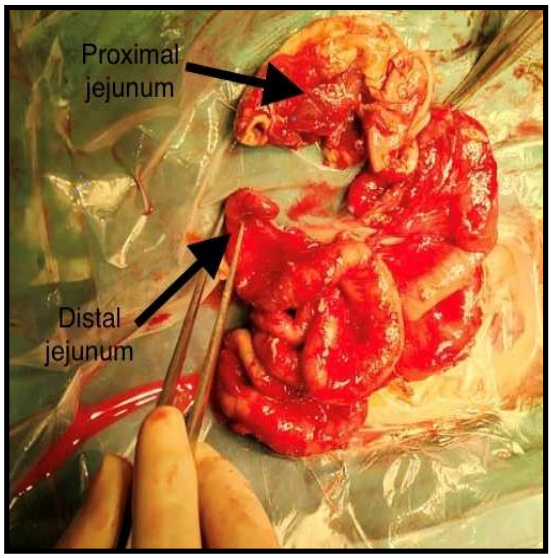

Figure 6 | Intraoperative photograph taken during laparotomy showed segmental jejunal volvulus in which the involved bowels are gangrenous and perforated. Distal jejunum was atretic (jejunal atresia type I).

Discussion

Unlikely jejunoileal atresia (JIA) which is rarely associated with other abnormalities, more than 50% of patients with duodenal atresia has other associated abnormalities such as Trisomy 21 (~50%), congenital cardiac defects (25-65%), malrotation (30%) and other gastrointestinal anomalies[2, 3]. The association of intestinal atresia with biliary atresia (BA) is even rarer and not well known. Based on the Japanese Biliary Atresia Registry from year 1989 to 2018, the incidence of intestinal atresia associated with biliary atresia was seen in 13 out of 721 of recorded cases, which is less than 2%[1].

In our cases, both of the patients were on long duration of TPN and their serum bilirubin levels were moderately elevated. Even though they were started on ursodeoxycholic acid to promote bile secretion and bile flow from the liver, there is no improvement in the liver function test. Since they were passing acholic stool, suspicion of BA was raised. In diagnosing BA, the gold standard are liver biopsy and cholangiogram.

Only the first patient was proceeded with percutaneous cholangiogram and liver biopsy showed pathognomonic features of BA. The parents of our second patient were not keen for further workup but the patient is highly suspicious of concurrent BA. Surgery involving resection of the atretic part of the intestine and primary anastomosis to restore significant bowel length are the principles in the management of intestinal atresia, while early Kasai portoenterostomy is crucial for native liver survival.

The presence of concurrent intestinal atresia and BA makes the Kasai operation more challenging and requires modification. Asabe et al. reported thirteen cases of BA with JIA which proceeded with Kasai operation, out of that only six patients survived to date, giving a survival rate of 46.2% [4]. Hans et al. reported five cases of meconium peritonitis secondary to perforation of small bowel atresia which later confirmed to have biliary atresia and proceeded with Kasai procedure. Two patients died of ascending cholangitis, one patient of liver failure that was exacerbated by TPN-associated liver injury, and one patient is awaiting a liver transplant. Only one patient is in good health, being anicteric, and showing normal growth and development[5].

Hess et al. reported a case of duodenal atresia with biliary atresia post Kasai portoenterostomy and currently awaiting liver transplant[6]. Chen et al. reported a case of successful modified Kasai operation whereby a segment of the colon is used as a biliary conduit for a case of small bowel volvulus with biliary atresia who had undergone extensive bowel resection[7].

For our patients, none of them proceed with Kasai portoenterostomy taking into consideration that the patients had undergone laparotomy and extensive bowel resection for intestinal atresia, the challenges of re-laparotomy and the relatively short bowel length remain post bowel resection and anastomosis.

Conclusion

Concurrent intestinal atresia with biliary atresia although uncommon, is important to take note of the possibility of these associations.

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