Case report

A RARE CASE OF DUODENAL ATRESIA WITH SITUS INVERSUS ABDOMINALIS

ABSTRACT

Background: Duodenal atresia is a cause of intestinal obstruction in the new-born and can be associated with other congenital anomalies such as intestinal malrotation and congenital heart diseases. Association of duodenal atresia with situs inversus abdominalis is extremely rare. It is characterised by bilious or non-bilious vomiting in 24 to 38 hours of neonatal life, typically following the first oral feeding.

Case report: A ten hour old male neonate delivered at 37weeks presented with bilious vomiting. There was polyhydramnios in third trimester. There was no abdominal distension and beyond 48hrs no meconium, systemic review was normal. On examination, he was febrile and tachypnoeic. Naso- gastric tube drained bilious effluent and bowel sounds were absent. He had a grade 3 systolic murmur at the left lower sternal border. A plain abdominal radiograph revealed a reverse double-bubble sign without dextrocardia. Echocardiography done revealed a congenital mitral incompetence. Based on these findings, diagnosis of duodenal atresia with situs inversus abdominalis and congenital heart disease was made. The child had a laparotomy with intraoperative findings of type 3 duodenal atresia, dilated stomach and proximal duodenal segment, complete situs inversus abdominalis, polysplenia, intestinal malrotation and midgut volvulus. He had Kimura diamond duodeno-duodenostomy, Ladd's procedure and a transgastric, transanastomotic tube was passed. He passed stool and breast feeding commenced via transgastric transanastomotic tube on third and fourth post-operative day respectively. He did well and was discharged home.

Conclusion

It is of utmost importance to look out for congenital anomalies when evaluating patients with duodenal atresia. This case reiterates the significance of reverse double bubble sign as an important preoperative means of recognizing this condition.

Key words: Duodenal atresia, reverse double-bubble sign, situs inversus abdominalis.

Introduction

Duodenal atresia is a common intestinal atresia treated by paediatrics surgeons and it occurs in 1: 5000-10,000 ~~livebirths~~live births. [1] Situs inversus is a rare condition with a frequency of about 1 in 10,000 of the normal population. [2] Duodenal atresia is associated with Down's syndrome (30%), annular pancreas (23%), congenital heart disease (22%), malrotation (20%), oesophageal atresia (8%), others (20%). [3] There is a rare association of duodenal atresia with situs inversus abdominalis, just about 20 cases have been reported in the literature so far. [4]

Case report

A ten hour old male neonate presented with ~~bilous~~bilious vomiting usually after feeding noticed since birth. He was delivered by caesarean section at 37 weeks gestation to a 38year old Para 3 mother and weighed 2.5kg at birth. An ultrasonography done at 36 weeks of gestation revealed polyhydramnios. The pregnancy was otherwise uneventful. His parents were non-consanguineous.

Physical examination revealed a pink baby saturating at 94% in room air. He was febrile with an axillary temperature of 37.8^{o}C and was not dehydrated. His abdomen was full with marked epigastric fullness and moved with respiration, there were no areas of tenderness and no palpable organomegaly. His bowel sounds were absent; his anus was patent and normally placed with good sphincteric tone. He had normal male external genitalia. The pulse rate was 140 beats per minute, which was regular and normal volume. He had normal 1st and 2nd heart sounds with a grade 3 systolic murmur heard at the left lower sternal border. He was tachypneic with a respiratory rate of 80 cycles per minute but had equal chest expansion and good air entry bilaterally.

Comment [D1]: THE IMPORTANT PRESENTING COMPLAINTS SHOULD COME BEFORE THE PHYSICAL EXAMINATION

Comment [D2]:

Comment [D3]: THESE SENTENCES SHOULD COME UP BEFORE THE ABDOMINAL EXAMINATIONS

He was placed on nil per oral, commenced on intravenous fluid for hydration at maintenance, was also placed on intravenous antibiotics and had a nasogastric tube passed for gastric decompression. 60ml of bilious effluent was obtained, hence the need for continuous nasogastric tube drainage. He had not pass meconium by the fifth day of life.

As part of work up for the clinical diagnosis of a congenital intestinal obstruction, he had a plain abdominal radiograph done which showed a reversed double bubble sign without dextrocardia (Figure 1). Based on this a diagnosis of duodenal atresia with situs inversus abdominalis was made. He also had an echocardiography done which revealed that the anterior mitral valve leaflet was elongated and prolapsed under the posterior leaflet. There was also minimal excursion of the posterior mitral leaflet and significant mitral incompetence with a jet length of about 2.4cm into the left atrium.

Following stabilisation and optimization he had a laparotomy done on the fifth day of life. The intraoperative findings (Figure 2, 3 & 4) were that of clear peritoneal fluid, dilated stomach and proximal duodenal segment, collapsed distal duodenum and small bowel (type 3 duodenal atresia), complete situs inversus abdominalis, intestinal malrotation, midgut

volvulus and polysplenia. He then had Kimura diamond shaped duodenoduodenostomy, release of Ladd's band, without appendectomy and a transgastric transanastomotic tube was passed.

Post operatively, he had nasogastric drainage until he passed stools on the 3rd day post-op, after which he was commenced feeds on the 4th day after surgery. He tolerated feeds and made a full recovery. He was discharged home and seen for follow up to assess the wound healing and adequacy of nutrition two weeks after discharge. He was then subsequently referred to the paediatric cardiologist for follow up.

Discussion

Duodenal atresia can take many forms, but proximal and distal intestinal segments always end blindly. [5] In the Type 3, the proximal and distal segments of the duodenum are completely separated as was found in the index case. [6] Duodenal obstructions can be complete or partial. Our index patient had a complete obstruction with a gap between the proximal and distal segments. Intrinsic duodenal obstructions may be attributed to atresia or mucosal webs. Duodenal extrinsic obstruction can occur in association with malrotation or a pre-duodenal portal vein. Hallmarks of duodenal atresia are a dilated proximal segment and a decompressed distal segment.

Duodenal obstruction is characterized by a double-bubble sign on prenatal ultrasonography. The first bubble corresponds to the stomach and the second to the post-pyloric dilated duodenum. [7] It is important to note that prenatal ultrasonography does not reliably detect duodenal atresia as was the case in the index patient. This is because in the presence of fetal vomiting, foetuses with duodenal atresia may have normal ultrasonographic findings. Prenatal diagnosis would have allowed for prenatal counselling of the mother and planning of management. Polyhydramnios was however, detected in the maternal antenatal ultrasonography of the index case. An absence of swallowing like in oesophageal atresia or a blockage of the foetus's gastrointestinal tract like in duodenal atresia and stenosis can lead to polyhydramnios.

Comment [D4]: DIFFICULTY IN SWALLOWING

Presenting symptoms and signs of duodenal atresia are the result of proximal intestinal obstruction. Duodenal atresia is typically characterized by the onset of vomiting within hours of birth as was seen in our index case. Gastric aspiration volume in a new born of more than 20 ml is suggestive of intestinal obstruction; normally, aspirates should be minimal. [8] The child in this report had as much as 60 ml of billous aspirate at the initial gastric decompression.

Upright plain abdominal radiograph of the new-borns which demonstrate double and distal bowel devoid of intestinal gas have diagnostic values. [9] This is how the diagnosis of duodenal atresia was made in our index patient. (Figure 1)

When literature series are reviewed, in more than 50% of the cases association of duodenal atresia with Down syndrome, oesophageal atresia, anal atresia and cardiac problems can be

seen. [3] The index case has congenital heart disease (congenital mitral incompetence), complete situs inversus abdominalis, intestinal malrotation/volvulus, and polysplenia.

Surgery is the definitive treatment for congenital duodenal atresia. However, adequate intravenous hydration, and gastric decompression are essential until the neonate is stabilized for surgical repair. Dehydration, weight loss and electrolyte imbalance soon follows if fluid and electrolyte losses are not adequately replaced. The patient being discussed had adequate fluid and electrolyte replacement preoperatively.

In patients with duodenal obstruction, a duodeno-duodenostomy is the most commonly performed procedure for operative repair. The patient being discussed in this report had an upper right transverse incision and Kimura diamond duodeno-duodenostomy. He had detorsion of volvulus anticlockwisely, ladd bands released, without appendectomy. Also to facilitate post-operative feeding, a transgastric transanastomotic feeding tube was passed. Studies have shown that placing a transanastomotic feeding tube has a beneficial effect on the time to full oral feeding. [10] Our index case was able to attained full oral feeding by the 8th day post-op.

Conclusion

Duodenal atresia is an uncommon cause of neonatal intestinal obstruction. Duodenal atresia association with situs inversus abdominalis is very rare, a high index of suspicion is necessary particularly when plain radiograph reveals a reverse double bubble sign. With advances in the neonatal intensive care, and refinement of surgical techniques there has been improvement in the survival rate over the years. Also prompt recognition of duodenal atresia with associated situs inversus abdominalis allows for referral to facilities with paediatric surgeons and also allows for preoperatively planning of surgical incision. In the absence of life threatening associated anomalies, the prognosis is good as was in this case.

References

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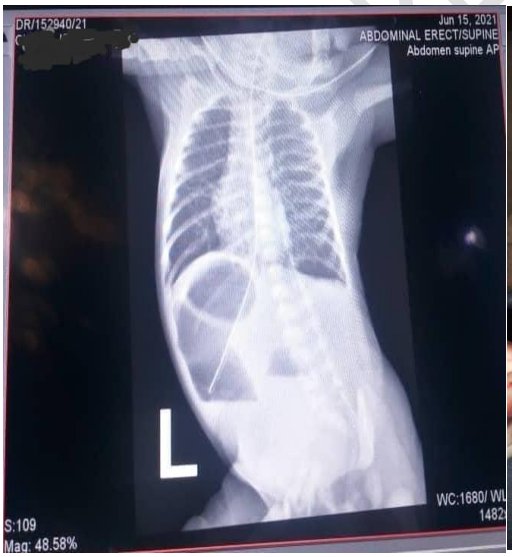

Figure 1: Reversed double bubble sign

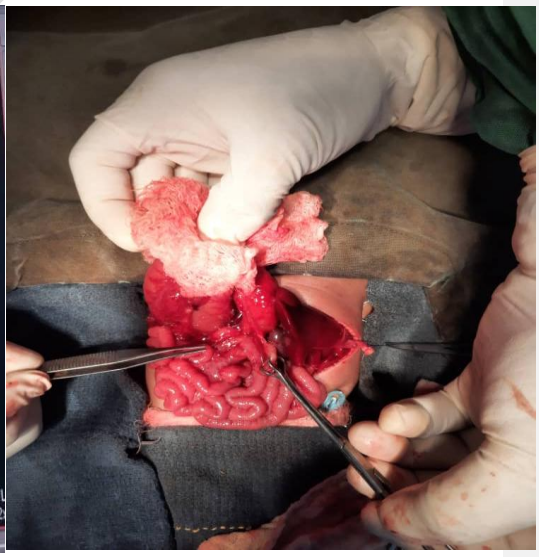

Fig 2. Type 3 Duodenal atresia

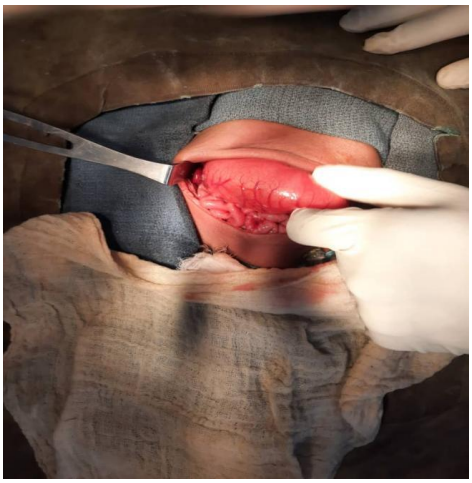

Fig 3. Poly splenia in the right hypochondria

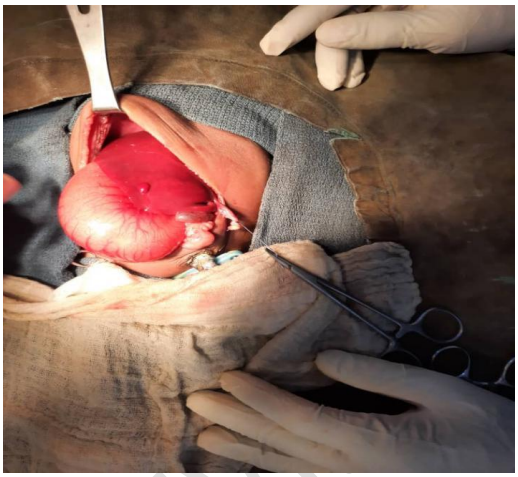

Fig 4. Liver in the left hypochondria