ALIMENTARY TRACT DUPLICATIONS: A REPORT OF SIX CASES

H. Pourang., V. Mehrabi and N.Ostadian

Department of Pediatric Surgery, Amir Kabir Hospital, Faculty of Medicine, Tehran University of Medical Sciences, Tehran, Iran

Abstract - Six cases of gastrointestinal duplications with varied manifestations are being presented. Case 1 presented with an epigastric mass, case 2 presented with an epigastric mass along with projectile vomiting since birth time, case 3 had severe colicky pain and was admitted as an emergency case, case 4 presented with massive rectal bleeding along with hypovolemic shock, case 5 was admitted for constipation of 12 years duration and case 6 was admitted as a suspected case of intussusception or Meckle's diverticula presenting with severe rectal bleeding and a hemoglobin of 6 g.
Acta Medica Iranica 36 (2): 79 - 86; 1998

Key words: Duplications, alimentary tract

INTRODUCTION

Duplications of gastrointestinal tract are rare congenital malformations that can occur from the mouth to the anus (1 - 8). According to some reports it can also occur at retroperitoneum and intrapancreatic regions (2 - 9). These malformations may be asymptomatic or depending on their type, size and location, various manifestations may be seen. Although improvement of imaging technology has made prenatal and postnatal diagnosis possible, but diagnosis is most accurate during operation (2 - 8). The aim of this paper is to introduce 6 cases of duplication which have been treated by us.

MATERIALS AND METHODS

Case 1

A 3 months old girl with an epigastric mobile mass (5×5 cm) was admitted to our hospital on July 29, 1994. Routine laboratory tests were normal. Barium meal, ultrasonography (US) and abdominal computerized tomography (CT) were performed. In barium study, the C-Loop of duodenum was slightly wide and in CT and US a cyst was seen in pancreatic head area (Fig. 1). With an initial diagnosis of mesenteric or pancreatic cyst, the child underwent elective laparotomy. There was a cyst about 7 × 6 cm in diameter along the pyloric canal, up to the first portion of duodenum. A strip of aberrant pancreatic tissue about 1cm wide and 7 cm long was seen upon the cyst (Fig. 2). The pancreas was small and abnormal, in its usual position. Separation of the cyst from pyloric canal and duodenum was not feasible. Therefore we resected the cyst and pyloric canal and a small portion of the beginning of duodenum near ampulla. Duodenum stump was closed and gastrojejunostomy was done. The postoperative period was uneventful and the infant was discharged 9 days after the operation. The pathologic report was cystic duplication of pyloric canal.

Case 2

On November 25, 1994, a 9 months old girl was admitted to our center with a history of projectile vomiting since birth. Vomiting was intermittent with periods of weeks without vomiting. A pediatrician had diagnosed her as a case of hypertrophic pyloric stenosis. On admission the general condition of the infant was good. In addition to vomiting, a non tender and mobile epigastric mass, about 6 cm in diameter was palpated. Barium study, ultrasonography and abdominal CT were done (Fig. 3). In US and CT, a cyst was seen near pancreatic head (Fig. 4). Like the previous patient with preoperative diagnosis of gastro - intestinal duplication, the infant underwent surgical exploration. On operation, a cyst with a diameter of 7 cm was found in pyloric region (Fig. 5). Separation of cyst from pyloric canal was not feasible. Therefore, complete excision of stenotic pyloric canal was done, gastrointestinal continuity was restored by gastroduodenostomy. There was no communication between the cyst and the severed stenotic canal of pylorus. The postoperative course was uneventful and the patient was discharged on the 5th postoperative

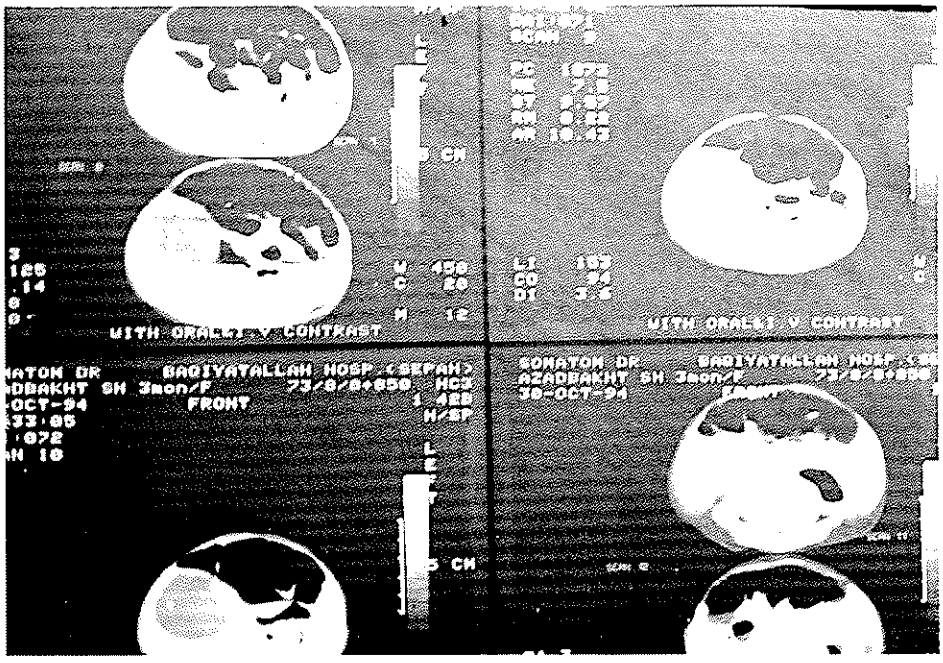

Fig. 1. Abdominal CT of case 1

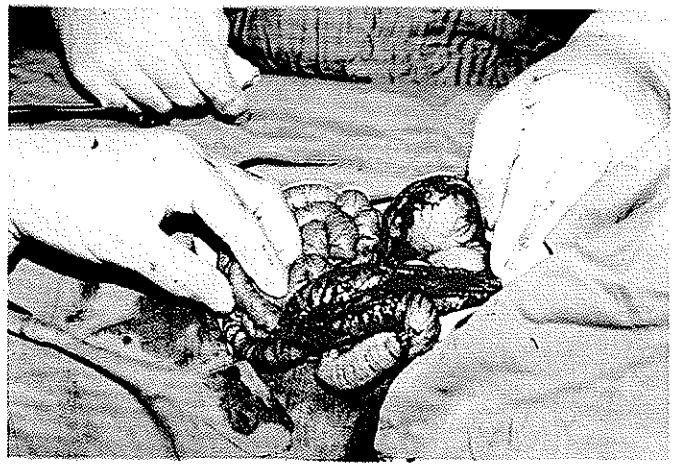

Fig. 2. Appearance of a pyloric cyst, case 1, note the aberrant pancreatic tissue on the cyst

day. Histopathologically the wall of the cyst consisted of gastric mucosa, circular and longitudinal smooth muscle layers and serosa.

Case 3

On November 16, 1994, an 8 year old boy was admitted to our hospital with a history of severe nocturnal epigastric pain. The epigastric pain became worse in supine position and was relieved by assuming an upright position and walking. The pain could not be subsided by a sedative or even morphine injection. Two years ago, he had a history of similar abdominal pain that had improved without any treatment. Physical findings, haematologic and biochemical parameters were normal except for serum amylase that was in the upper normal limit. An upper gastrointestinal (UGI) series ultrasonography and abdominal CT was performed. In UGI series, duodenal C - loop was slightly wide and in US and CT a small cyst, about 4 cm in diameter was seen around pancreatic head. Due to the continuation of severe abdomianl pain, the boy underwent exploratory laparotomy. The lesser sac was opened, pancreas was large, firm with nodularity (chronic pancreatitis). Kogher manoeuvre was performed and with compression of duodenal loop a cyst of about 5cm in diameter was seen and palpated (Fig. 6). The location of the cyst was intraduodenal, posterolateral to the second portion of the duodenum. The cyst contained a cloudy fluid on aspiration (Fig. 7). As the cyst was near the ampulla, excision was not feasible and hence internal drainage (transduodenal cystoduodenostomy) was performed. After drainage, the cyst wall collapsed. There was no communication between the cyst and duodenal lumen. The postoperative course was uneventful except for severe nocturnal pain, only during the first postoperative night. We checked the boy 5 months after the operation. He did not experience any abdominal pain. The pathological finding was duplication of duodenum with ectopic gastric mucosa in the cyst.

Case 4

A two year old girl was referred to us in February 1992 with massive rectal bleeding and shock. Rectal bleeding was painless and had started 3 days earlier. In the previous year she had been admitted to another hospital for rectal bleeding. After blood transfusion the rectal bleeding had stopped spontaneously and the child had been discharged. In our centre, rectal bleeding continued despite 3 blood transfusions. Therefore, we performed only routine laboratory tests and emergency laparotomy. Our preoperative diagnosis was Meckle's diverticulum and we found a tubular duplication about 25 cm long, 70 cm from ileocecal valve. The duplication was in mesenteric position with a blind proximal end and an open distal end that was communicating to normal ileal lumen. The distal end was red, firm and haemorrhagic (Fig. 8). Because the blood supply of the duplicate and the normal ileum was common, resection of the normal bowel and duplicated portion was followed by an end to end anastomosis. The postoperative period was excellent and the child was discharged on the 5th post operative day. Pathologic finding was tubular duplication of ileum with ulceration due to ectopic gastric mucosa.

Case 5

A 12 year old girl was referred to us in 1983 for evaluation of chronic constipation. The constipation had started at birth. She was treated for some years for habitual constipation. With huge megacolon on barium enema and an initial diagnosis of Hirschsprung's disease, the patient underwent rectal and abdominal biopsy (Fig. 9). Pathologic examination showed normal ganglion cell in colon wall. Two months after the second biopsy, the child underwent a second laparotomy. On careful exploration, we found a huge duplicated megacolon along the left side, that communicated to adjacent proximal and distal ends of bowel. Complete resection of duplicated and normal sigmoid and an end to end anastomosis of left colon was done. Post operative course was without complication and the child was discharged 10 days after the operation. Subsequent follow up showed no constipation until 3 years after the surgery.The pathologic finding was tubular duplication of sigmoid colon.

Case 6

An 8 month old boy was admitted to our hospital on

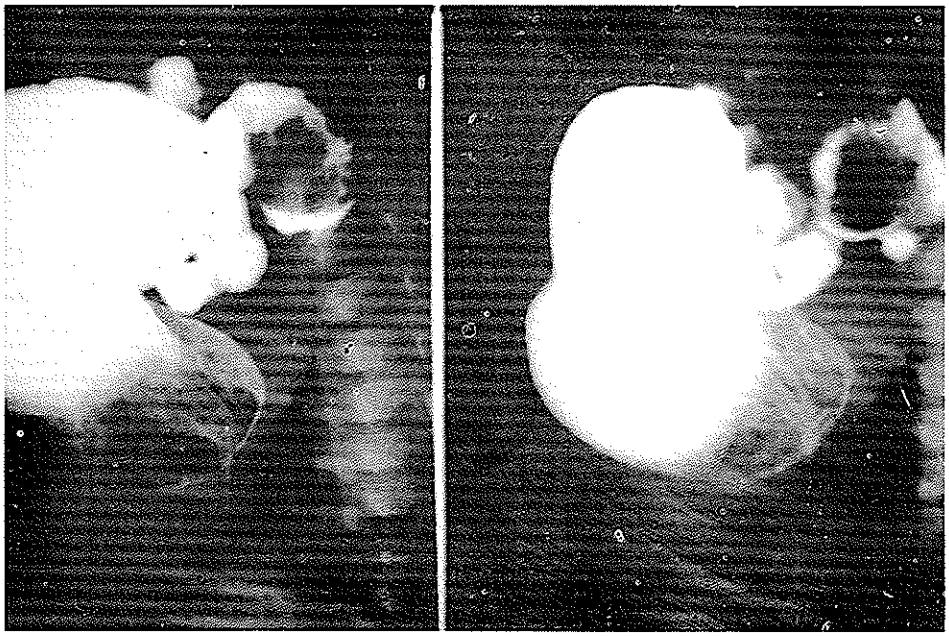

Fig. 3. Barium study of case 2

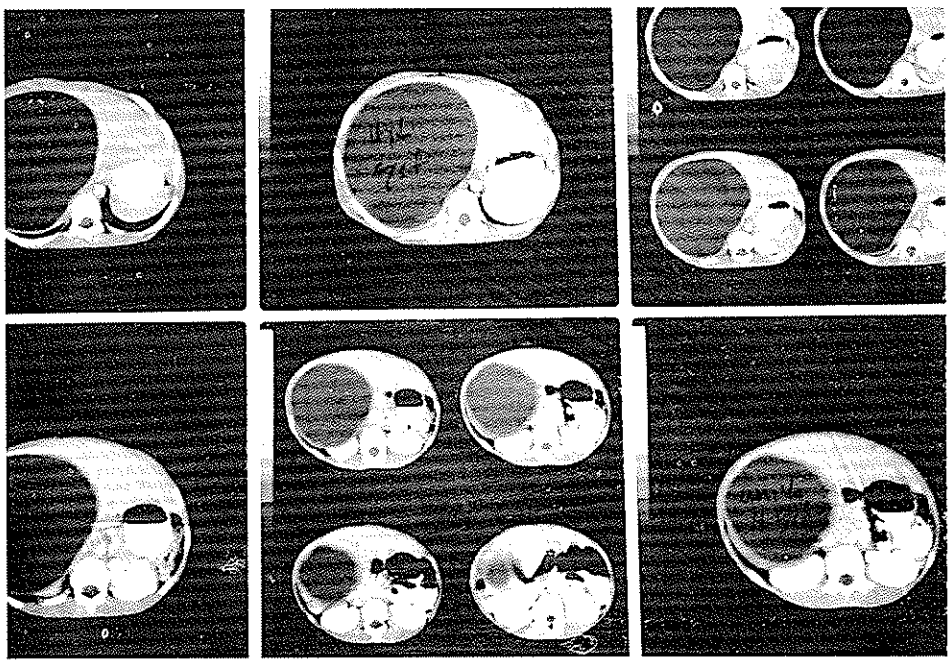

Fig. 4. Abdomial CT of case 2

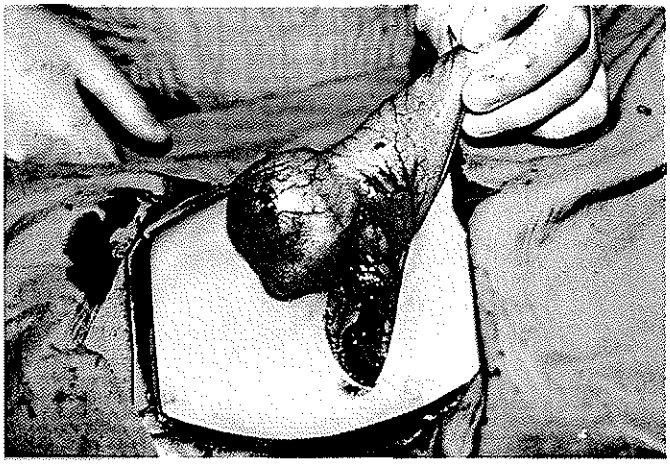

Fig. 5. Operative photograph demonstrating pyloric canal duplication

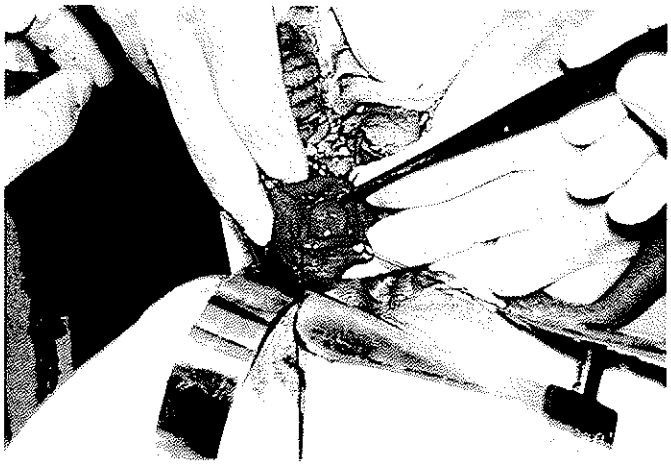

Fig. 6. Appearance of intraduodenal cyst (case 3) after doudenotomy

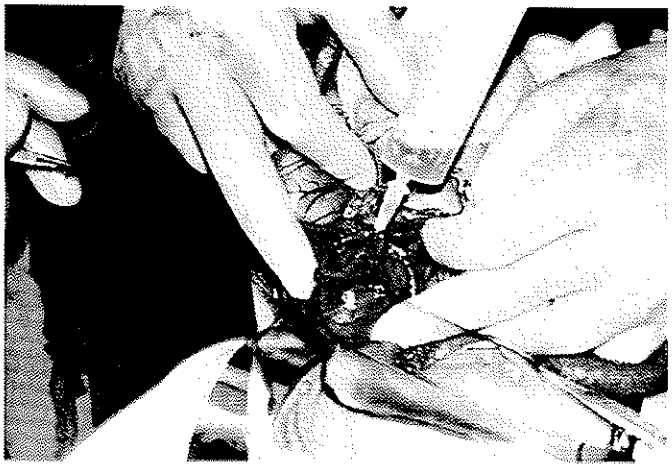

Fig. 7. Aspiration of intraduodenal cyst content

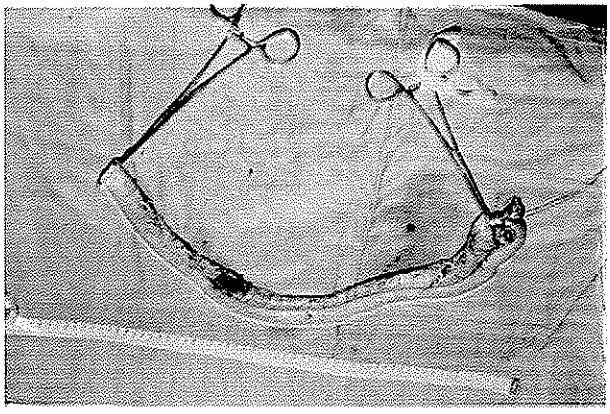

Fig. 8. Operative photograph of tubular duplication of ileum. Note the haemorrhagic site of ileum due to ectopic gastric mucosa

March 3 1997 with massive rectal bleeding (hemoglobin = 6 g/dl) and generalized abdomoinal tenderness. Because the patient was in shock with continuous bleeding, he underwent an emergency laparotomy with primary diagnosis of intussusception or Meckle's diverticula. During the operation, it became clear that the source of bleeding was a 10 cm tubular duplication of terminal ileum. A resection of the duplicated area and end to end anastomosis was performed. The patient was discharged 6 days after the operation in a good general state. The pathologic report stated a tubular duplication of ileum with ectopic gastric mucosa.

DISCUSSION

Duplications of the alimentary tract are rare congenital malformations (1 - 8) that consist of cystic or tubular structures lined by normal gastrointestinal mucosa with a smooth muscle wall like that of the gastrointestinal tract, that occur in proximity to all parts of alimentary tract from mouth to anus and according to some papers in retroperitoneal or intrapancreatic areas (2 - 9). There are some theories about the origin of duplications such as partial twinning, residue of the neuroenteric canal, embryonic diverticula and abnormal recanalization of the intestinal lumen after solid stage of embryologic development (8). Duplications are cystic (90%) or tubular (10%). The usual location is on the mesenteric aspect of bowel, in contrast to Meckle's diverticulum with common blood supply. The most common location of duplication is in jujeno-ileal region (50%). The signs and symptoms of these malformations depend on their size, shape and location. In some cases there are no symptoms. Mass, vomiting, intestinal obstruction (due to compression, volvulus, intussusception) abdominal pain (peptic ulcer or pancreatitis), anaemia and peritonitis. (7 - 8) are

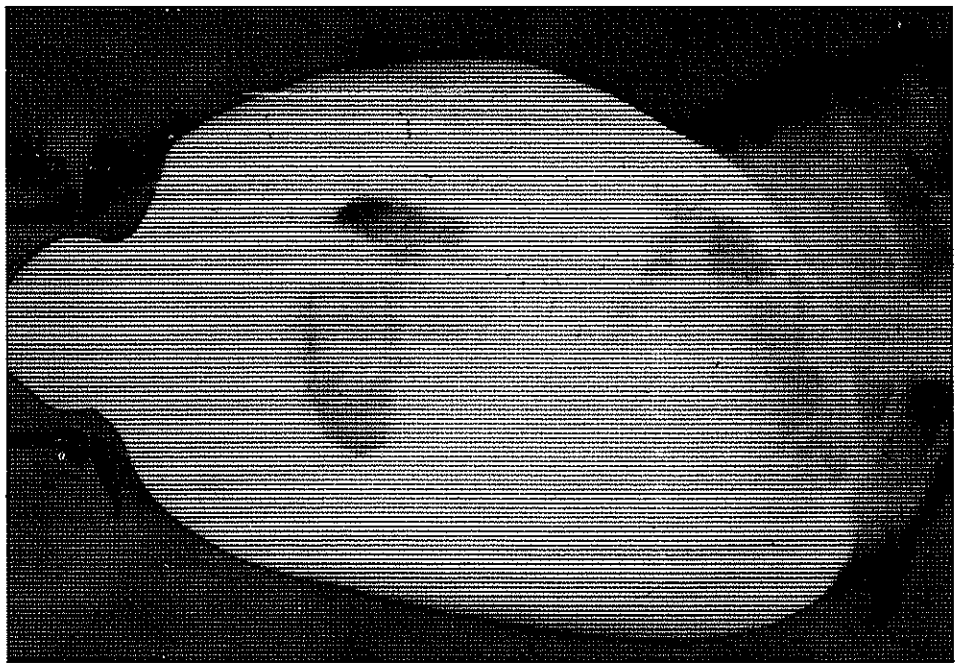

Fig. 9. Barium enema case 5. Note the megacolon due to duplication of left colon

the most common findings. Duplications of the gastrointestinal tract are often associated with other congenital malformations such as vertebral lesions (hemivertebra) omphalocele, intestinal atresia or stenosis and malformation of mid - gut. With advanced imaging technology, the duplication can be detected prenatally, but accurate diagnosis is only feasible at operation. Complete removal of small duplicated cysts and end to end anastomosis in tubular form are ideal, but in large cycts or long tubular duplications of more that 50 cm, these techniques are not feasible and there are many other approaches in these instances such as partial excision, internal drainage and stripping of mucosa of duplicated segment (4, 5, 8). There is a new experience in long tubular duplication that the authors have separated the duplicated portion from normal adjacent bowel with preservation of common vessels. We had no duplications in thorax. Case one and two had cystic duplications of pyloric canal. The incidence of duplication in this region are 3.8 to 5.2 percent (4). In case 1, an epigastric mass was the only symptom but in case 2, projectile vomiting and epigastric mass were the chief complaints. Separation of the duplication cysts from the pyloric canal was not feasible, therefore complete resection of cyst and pyloric canal and gastrojejunostomy in case 1 and gastroduodenostomy in case 2 were performed. In case 1, the distal margin of the cyst was in close proximity to ampulla with a strip of aberrant pancreas upon it. The third case had chronic recurrent pancreatitis with severe abdominal pain at night. The surgical finding was posterolateral intraduodenal duplication cyst. Duplication of the alimentary tract causing pancreatitis is extremely rare and only 14 cases were reported till 1992 (10). The location of intraduodenal cyst is posteromedial, in contrast to our case. Intermittent compression of ampulla by the cyst probably was the cause of reflux pancreatitis in this case (6). After internal drainage and collapse of the cyst wall, the patient's signs disappeared. There was ectopic gastric mucosa in the cyst, therefore, in this case and two previous cases bile reflux gastritis and peptic ulceration may have occurred and these patients must be under close observation. In cases 4 and 6 with tubular ileal duplication, resection of the duplicated portion and the adjacent bowel was done. The source of rectal bleeding in these cases was peptic ulceration of the ectopic gastric mucosa in the duplicated bowel. Case 5, a 12 year old girl with left colon duplication was managed for many years with a mistaken diagnosis of constipation due to Hirschprung's disease. Resection of the huge duplicated segment and adjacent colon terminated 12 years of constipation in this patient. In conclusion 6 cases of rare GIT duplication from gastroduodenal canal to sigmoid area are described and discussed. These cases had bizarre manifestations and had caused problems in diagnosis and treatment. Misdiagnosis could eventually result in the death of the patients. Considering the variations of GIT duplication would be beneficial in timely diagnosis.

REFERENCES

1. Cigliano B. Ascione J. Ped. Surg. Sci. 8: 27 - 29; 1994.

2. Duncan N. and Adzick Sciut. Retroperitoneal alimentary tract duplication detected in utero. J. Paediatric Surg. 27: 123 - 133; 1992.

3. Balen J. F. and Hernandes L. J. Pediatric Surg. 28: 1588 - 1594; 1993.

4. Murty T.V.M. and Bhargva R.K. J. Pediatric Surg. 27: 515 - 517; 1992.

5. Nokrris R.W. and Brecton R.J. 21: 167; 1986.

6. Okuyma H. and Matso Y. Intrapancreatic duodenal duplication associated with pancreatic pseudocysts. J. Paediatric Surg. 27: 1573 - 1574; 1992.

7. Role G. and Doing M. Perforation of the jejunum secondary to duplication cyst lined with the ectopic gastric mucosa. J. Paediatric Surg. 23: 1025 - 1026; 1988.

8. Wrennearle L. Alimentary tract duplication. Ashcraft Holder Paediatric Surg. W.B. Saunders Co. 1993: 421 - 433; 266 - 271.

9. Wold M. and Callery M. Ectopic gastric - like duplication of the pancreas J. Paediatric Surg. 23: 1051 - 1052; 1988.

10. Black Preston R. and Welch Kenneth J. Juxtapancreatic intestinal duplications with pancreatic ductal communication, a case of pancreatitis and recurrent abdominal pain in childhood. Paediatric Surg. 21: 257 - 261; 1986.

11. Bergmann K.S. and Jacir NN. J. Cytic duodenal duplication: Staged managment in a premature infant. Pediatric Surg. 28: 1584 - 1585; 1993.