# Intestinal perforation by multiple ectopic pancreatic tissues in a neonate with multiple congenital anomalies

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## ABSTRACT

يتم تشخيص النسيج البنكرياسي المنزاح في الجهاز الهضمي بشكل عرضي عن طريق جراحة البطن أو تشريح الجثة. ومن النادر وجود أعراض مصاحبة لهذه الأنسجة المنزاحة كالنزف، الالتهاب البنكرياسي، انسداد أو إنثقاب الأمعاء. في هذا الملخص تُعرض حالة خديج ذو تشوهات خلقيِّة متعددة بالإضافة لوجود تعدّد الأنسجة البنكرياسية المنزاحة في الجهاز الهضمي مسببة لنزيف وثقب بالأمعاء.

Ectopic pancreatic tissues of the gut are usually found incidentally during laparotomy or are reported in the autopsy findings. Rarely these ectopic pancreatic tissues may cause symptoms such as hemorrhage, pancreatitis, intussusception or perforation. We present a case report of the presence of multiple ectopic pancreatic tissues in the gut causing hemorrhage and perforation in a preterm, extremely low birth weight neonate with multiple congenital anomalies.

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Ectopic pancreas is an aberrant tissue that occurs in the gastrointestinal (GI) tract, which usually is an incidental finding at autopsy or laparotomy. However, specific clinical manifestations can occur. We report a case of multiple ectopic pancreatic tissues complicated by hemorrhage and perforation of the gut in a preterm newborn associated with multiple congenital anomalies. We also discuss the embryopathology and management options of ectopic pancreatic tissues. Symptomatic ectopic pancreatic tissues have been sparingly reported in adults. We are reporting this as the first case report of symptomatic ectopic pancreatic tissues in a newborn in English literature.

**Case Report.** A preterm (34 weeks), extremely low birth weight female baby (1.3 kgs) was born to gravida 8 para 7 Saudi mother by cesarian section due to antepartum hemorrhage and fetal distress. There was positive history of polyhydramnios on antenatal ultrasound (USG) and blood stained liquor. The baby was intubated following birth due to severe respiratory distress and shifted to the neonatal intensive care unit (NICU). Apgar scores were 3, 5, 6, and 6 at 1st, 5th, 10th, and 15th minutes. Physical examination revealed facial dysmorphism in the form of low set ears, depressed nasal bridge, short neck, prominent occiput, narrow palpebral fissure, and micrognathia. A feeding tube failed to pass beyond 10 cms, and x-ray revealed esophageal atresia and tracheoesophageal fistula (type 3b). Auscultation revealed bilateral crepitations, and grade II/VI murmur over the left lateral sternal border. The abdomen was mildly distended with no organomegaly, Back, spine, limbs, and genitalia were normal. Echocardiography revealed atrial septal defect (ASD), bicuspid aortic valve, large patent ductus arteriosus (PDA) with bidirectional shunting. Abdominal USG showed an increase in echogenicity of the right kidney. Blood counts, and biochemical parameters were within normal limits. The baby was managed with high frequency ventilation, surfactant therapy, upper esophageal pouch suctioning, and inotropes for hypotension. On the 4th day of life, the baby developed abdominal distension and pallor. Hematocrit had dropped from 36-28%, and x-ray of the abdomen revealed gastric distension, with no free abdominal air. A decision to place a gastrostomy tube for decompression was taken. At laparotomy, hemorrhagic bile stained ascites was found, and there were multiple polypoid tissues at the antimesenteric side of the jejunum and ileum (at least 4 in number, and each measuring 3-5 mm in size, **Figures 1a & 1b**). There was a perforation of the proximal jejunum (25 cms from the duodeno-jejunal [DJ] junction) adjacent to the abnormal tissue. Resection of a segment of jejunum (4 cms) bearing perforation and abnormal tissue, end-

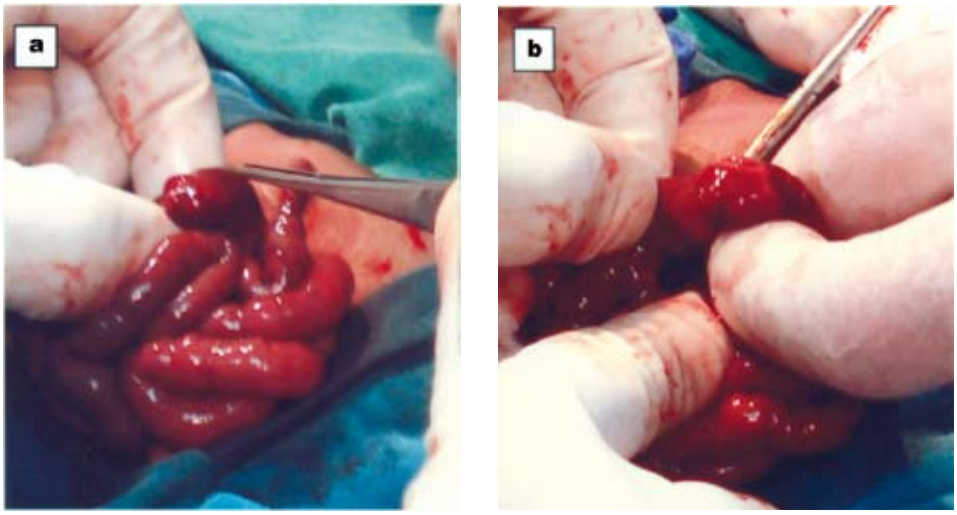


**Figure 1 -** Polypoid tissue at the **a**) antimesenteric border of jejunum with a small perforation, **b**) proximal ileum.

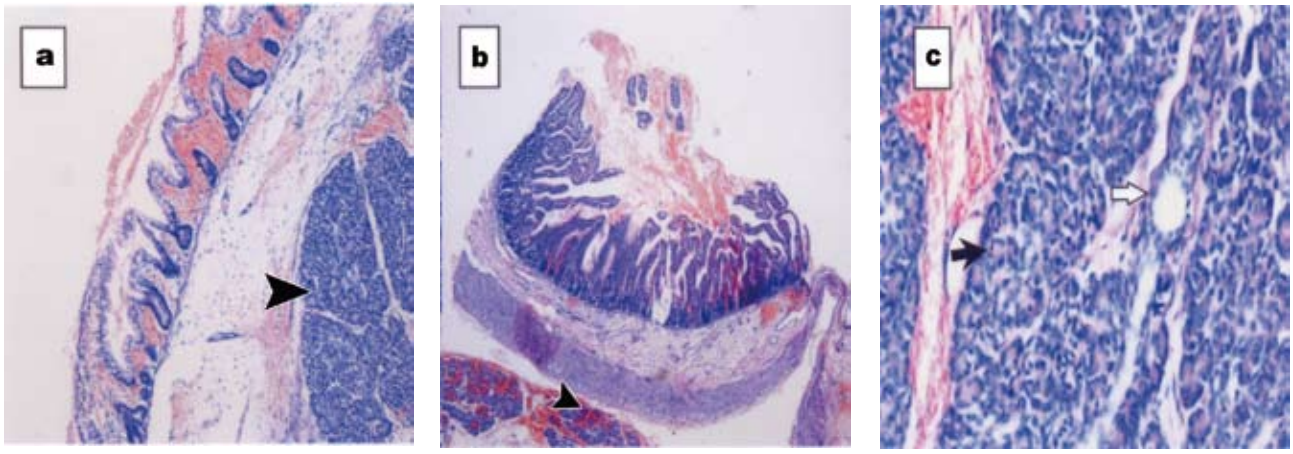


**Figure 2 -** Histopathology showing **a**) ectopic pancreatic tissue in the submucosa of jejunum, **b**) invasion into muscularis layer. **c**) Presence of acini (bold arrow) and few dilated ductal elements (block arrow) with absence of islets cells suggesting Heinrich's Type II ectopic pancreatic tissue.

to-end anastomosis, and Stamm's gastrostomy were made. Post operatively, the baby remained critical and finally succumbed to respiratory failure, hypotension, and *Staphylococcus* sepsis on the 24th day of life. Chromosomal analysis was reported to be normal. Histopathology revealed ectopic pancreatic tissue in the submucosa of jejunum invading into the muscularis (**Figures 2a, 2b, & 2c**). The ectopic pancreatic tissue was composed of acini and ductal elements without distribution of islets (Heinrich's type II).[1]

**Discussion.** Pancreatic ectopy (synonymous heterotopic, accessory, aberrant pancreas or pancreatic choristoma, and so forth) is defined as the presence of pancreatic tissue lacking anatomic and vascular continuity with the main body of the gland.[2-4] It can be found anywhere along the foregut and proximal midgut and usually causes no symptoms.[2-4] Approximately, 75% of all pancreatic rests are located in the stomach, duodenum, or jejunum.[2] However, they have also been found in the ileum. Meckel's diverticulum, gallbladder, common bile duct, splenic hilum, umbilicus, lung, and in perigastric and peri duodenal tissue.[3] In autopsy series, the frequency of ectopic pancreas is between 1-2% (range: 0.55-13%). The rate of recognition at the time of laparotomy is 0.2%. The normal pancreas develops from 2 primordial diverticula of the duodenum and becomes apparent at the 4th week of gestation.[4] After axial rotation of the gut, the mesoduodenum fuses with the posterior peritoneum and forms the most part of the duodenum. Fusion of the 2 pancreatic buds occurs simultaneously. During pancreatic organogenesis,

a number of embryological "mistakes" can happen. Pancreatic developmental errors can be classified as failures of migration or fusion and ductal anomalies.[5] Ectopic pancreatic rests arise from one or a combination of these developmental anomalies. Multiple ventral pancreatic buds may develop and their failure to atrophy and subsequent growth, and sequestration may give rise to ectopic pancreas. Parker[4] suggested that adhesion of portions of the dorsal and ventral pancreatic primordia to the neighboring structures during elongation and rotation of the gut may give rise to ectopic pancreas. Alternatively, budding of pancreatic tissue from the embryonic anlagen with attachment to the gut wall and subsequent separation may causes ectopic pancreas as well. It has been suggested that multipotential cells from the primitive gut, capable of specialized differentiation may form pancreatic rests.[4] Because of the close development of the embryonic pancreatic primordial buds to the foregut, it is not surprising that 70-90% of pancreatic ectopia occurs in the upper GI system.[6] Most ectopic pancreas in the upper GI tract are found in the gastric antrum: 75% are in submucosa, 15% in the muscular layers, and 10% in the subserosa.[7] However, heterotopic pancreatic tissue may be implanted extraintestinally, for example, gallbladder, common bile duct, liver, spleen, omentum, lungs, umbilicus, mediastinum, and so forth. In most cases, this anomaly is an incidental finding without requiring further investigations or management. The most common clinical symptoms attributed to ectopic pancreas are abdominal (epigastric pain), dyspepsia, and GI bleeding.[8] Pyloric obstruction by ectopic tissue, obstruction of the ampulla of Vater, the intestine and the biliary tree were described as well. Pancreatitis in the ectopic tissue, cyst formation, and inflammation with necrosis of the adjacent structures has been reported.[2,3,8] Several cases of cancer occurring in ectopic pancreas have been already described.[9] The GI bleeding can be attributed to ulceration of the overlying mucosa and disruption of submucosal vessels. Among the cases of ectopic pancreas with GI bleeding, the most common sites were the duodenum (26%), the stomach (21%), and the Meckel's diverticulum (21%).[10] The Jejunum (16%), ileum (11%), and the esophagus (5%) account for lesser incidences. Due to esophageal atresia and tracheoesophageal fistula associated with our case, there were no hematemesis and the proximal hemorrhagic contents of the gut were found in the lumen of the small intestine and partly mixed with extraintestinal bile subsequent to perforation. The event of bleeding into the gut lumen, known as hemorrhagia intestinalis is evident from the sudden drop in hematocrit and possibly the raised luminal pressure could have contributed to the perforation of the gut. Transmural perforation of gut, presence of multiple ectopic pancreatic tissues in jejunum and ileum and associated multiple congenital anomalies is also unique to our case. There have been no previous reports on any syndromic or nonsyndromic association of multiple pancreatic ectopy in neonates. The presence of Heinrich type 2 ectopic pancreatic tissue in the submucosa and muscularis layer, mucosal ulceration, and disruption of submucosal vessels due to pancreatic enzymes might explain the cause of hemorrhage and transmural perforation in our case as described in previous case reports.[9,10]

Management of ectopic pancreas is somewhat controversial. As this anomaly is largely asymptomatic and rarely associated with clinical pathology, the incidental finding of characteristic lesion does not necessarily warrant excision. Operative treatment is reserved for complicated cases, when clinical symptoms are associated with its presence such as recurrent bleeding, obstruction, or malignant degeneration, or when its appearance is difficult to differentiate from the other lesions such as leiomyoma or carcinoid. We decided to resect the segment of jejunum carrying the ectopic rest and perforation, and left behind the rest of the jejuno ileal pancreatic rests due to the poor general condition of the neonate. However, we recommend resection of all the segments of gut carrying these polypoid tissues and end-to-end anastomosis to avoid further complications of hemorrhage and perforation, especially when one of those has already caused complications at such an early neonatal age.

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