cases from the Cooky Jar
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Heterotopic Pancreatitis

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Heterotopic pancreas is a rare congenital condition in which pancreatic tissue deposits abnormally outside the pancreas (1). Heterotopic pancreas is a migration-type congenital anomaly (1). Although the embryologic mechanism is still unclear, three prevailing theories have been proposed: the misplacement, metaplasia, and totipotent cell theories (1–3). The most widely accepted theory, misplacement, suggests that pancreatic tissue fragments are deposited separately from the main gland during foregut rotation in the developing gastrointestinal system (1,2). Alternatively, the metaplasia theory suggests that endodermal cells migrate to the submucosa and transform into pancreatic tissue, and the totipotent cell theory proposes that intestinal endodermal cells differentiate into pancreatic tissue (1,2). Molecular and genetic abnormalities have also been postulated (3).

Heterotopic pancreas is usually discovered incidentally and is typically asymptomatic. However, it may develop any of the complications seen with the orthotopic pancreas. Acute pancreatitis is not always visible clinically or radiologically in the setting of heterotopic pancreas (Fig 1). Typically,

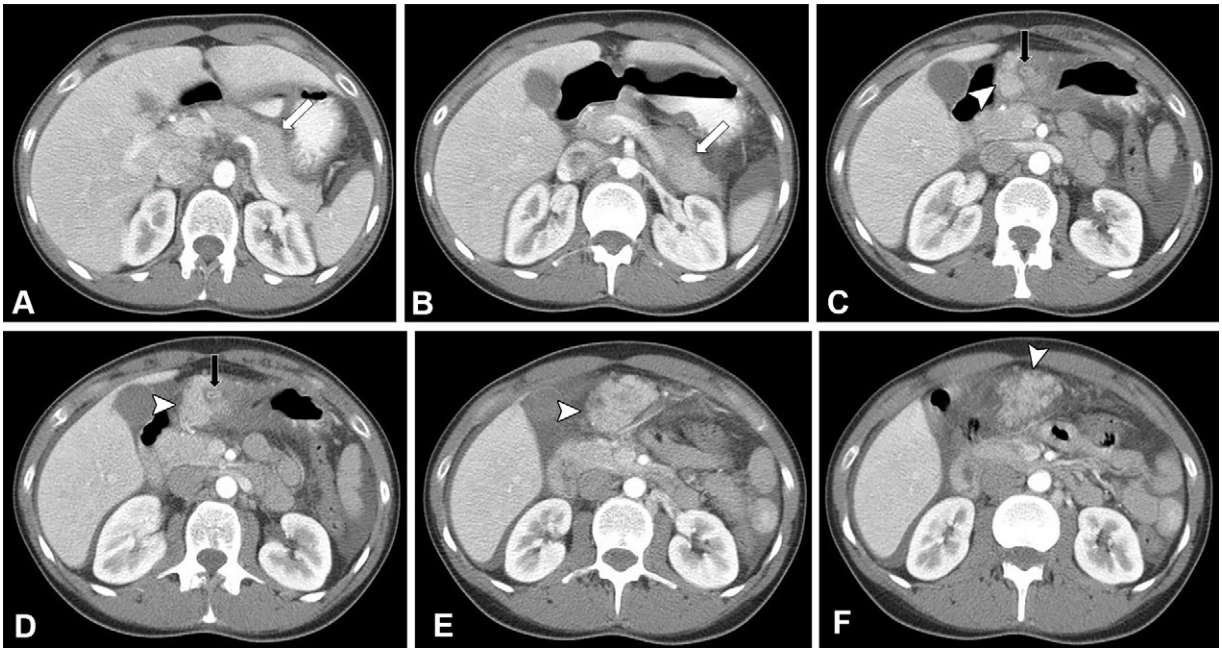

Figure 1. Heterotopic pancreas complicated by acute pancreatitis in a 28-year-old woman with abdominal pain and nausea (elevated serum lipase level of 1300 U/L). Axial contrast-enhanced CT images show the normal orthotopic pancreas without evidence of inflammation (white arrow in **A**, **B**). A lobulated mass is seen extending off the gastric antrum with a similar appearance to and enhancement as the orthotopic pancreas (arrowhead in **C–F**), with surrounding fat stranding. Note also a primordial ductlike structure centrally (black arrow in **C**, **D**). The most common CT appearance of heterotopic pancreas is an intramural mass with ill-defined microlobulated margins. The attenuation and enhancement characteristics are related to the histologic composition: acinus-dominant lesions demonstrate avid homogeneous enhancement, whereas duct-dominant lesions are hypovascular and heterogeneous after contrast agent administration.

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there is only mild enzyme elevation due to the small volume of inflamed tissues, and tissue may enhance less avidly when acutely inflamed (1,4). The location of the heterotopic tissue will dictate the site-specific complications and imaging features (Fig 2).

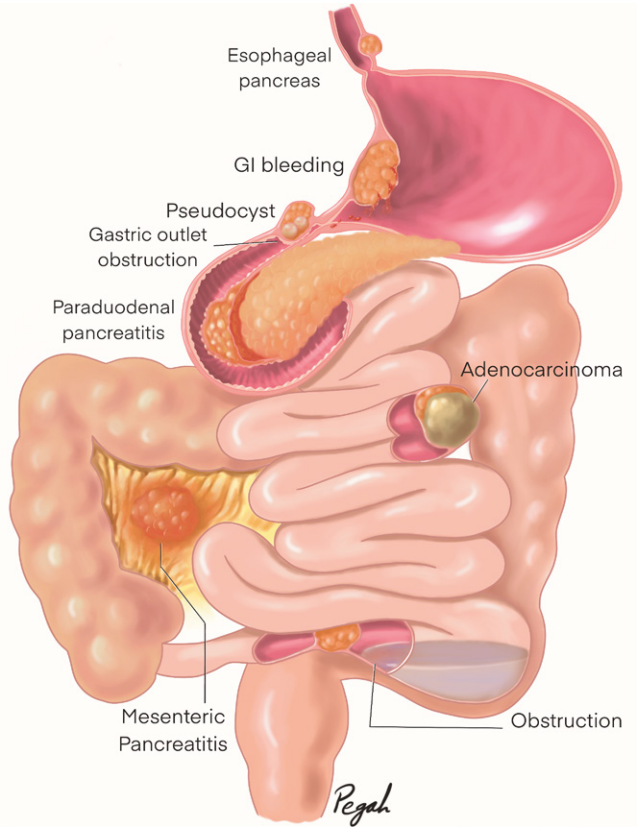

Figure 2. Medical illustration shows the most common sites of involvement and possible associated complications of heterotopic pancreas. Sites where heterotopic pancreas can be found include the gastrointestinal tract, Meckel diverticulum, mesentery, hepatobiliary system, and spleen, as well as other less common extragastrointestinal locations such as the mediastinum, lungs, fallopian tubes, umbilicus, and omentum (1). Common gastrointestinal locations of heterotopic pancreas include the stomach, duodenum, and proximal jejunum, possibly due to their shared origins from the primitive foregut (1,5). As in orthotopic pancreas, heterotopic pancreas is susceptible to pancreatitis, pseudocyst formation, and pancreatic neoplasms (1,2). Location-related complications include gastrointestinal bleeding, bowel obstruction, and intussusception (1). Paraduodenal pancreatitis likely results from deposited heterotopic pancreas tissue in the medial duodenal wall leading to abdominal pain and duodenal obstruction and can act as a lead point causing intussusception (1,6). Esophageal and gastric lesions may manifest with dysphagia, epigastric pain, gastroesophageal reflux disease, and possible hemoptysis (6). *GI* = gastrointestinal.

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