ERCP and EUS technique in situs inversus totalis: preparing for a left-sided plot twist

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BACKGROUND

Situs inversus totalis (SIT) is the rare occurrence of a mirror image reversal of the entire internal visceral anatomy with a reported incidence of 1 out of 8000 to 1 out of 25,000.1,2 The altered anatomy in SIT imposes significant challenges to performing diagnostic and therapeutic endoscopy successfully. We describe in this case report a successful EUS and ERCP in a patient with SIT.

CASE REPORT

A 64-year-old woman with a known history of SIT associated with complex repaired congenital cardiac abnormalities presented with a few days of worsening, dull, right lower-quadrant abdominal pain and a week of acholic stools, jaundiced skin, and diffuse pruritis. Physical examination was notable for significant jaundice and mild bilateral lower quadrant tenderness. Blood tests revealed elevated aspartate aminotransferase (197 U/L), alanine aminotransferase (141 U/L), alkaline phosphatase (814 U/L), and total bilirubin (28.3 mg/dL). A CT scan of the abdomen revealed moderate intrahepatic biliary dilation with a nondistended common bile duct and a heterogeneously enhancing gallbladder wall thickening with a small amount of wall calcification (Figs. 1 and 2).

Video description

The patient was placed in the left lateral position. First, we performed an EGD to help identify the altered anatomy. Afterward, a curvilinear echoendoscope was advanced by performing endoscopy techniques in the usual order but in inverse direction. In particular, the postbulbar examination required a counterclockwise rotation with the left arm hanging straight down to facilitate the visualization of the mesenteric vessels, uncinate part of the pancreas, ampulla, and the ductal anatomy.

Endoscopic ultrasound showed diffuse circumferential common bile duct wall thickening and a stricture extending from the ampulla to the hepatic bifurcation. The pancreatic duct, head, body, and tail were unremarkable (Fig. 3).

The patient was maintained in the left lateral position and ERCP was subsequently performed, as shown in Video 1 (available online at www.giejournal.org). Given the reversed anatomy, the ampulla of Vater was identified at 2 o’clock compared to patients with regular anatomy, where it is found at 10 to 11 o’clock (Fig. 4). A cholangiogram confirmed a single diffuse stenosis extending from the ampulla to the bifurcation. After biliary sphincterotomy, intraductal brushings were obtained, and a temporary biliary stent was placed (Figs. 5 and 6).

Outcomes

There were no immediate adverse events from the procedure, including post-ERCP bleeding or acute pancreatitis. Microscopic examination of bile duct brushings confirmed the diagnosis of adenocarcinoma.

DISCUSSION

There are limited reports in the literature of successful ERCP and even fewer of EUS in patients with SIT, as summarized in Tables 1 and 2. The mirror image technique was described, which parallels the inversed anatomy through the performance of all regular endoscopic maneuvers inversely.3 Similar to our method, a commonly cited technique relies on inserting the endoscope with a 180-degree clockwise rotation in the stomach or duodenum or both. However, the ease of insertion may come with difficulty cannulating the papilla, which usually sits in the 1 to 3 o’clock position.4-19 Another technique involved changing the patient position during the procedure, which brought challenges to the endoscopy and was limited by patient intolerance.20,21 There are few reports in the literature of successful EUS in patients with SIT. These case reports make note of challenges identifying vascular anatomy and suboptimal views of the pancreas requiring the use of multiple endoscopes or even changing the patient’s position.22-24

CONCLUSION

Situs inversus totalis is a rare congenital disorder that poses significant challenges when performing diagnostic and therapeutic endoscopy. Performing the EGD is not challenging in SIT, but colonoscopy, EUS, and ERCP can be more difficult than usual given the altered anatomy. Starting with...
an EGD is reasonable to ascertain the “lay of the land” before performing EUS or ERCP. An experienced endosonographer with good knowledge of anatomy can safely and successfully perform linear EUS-guided interventions. In addition, ERCP can be safely and successfully performed with largely “normal” patient and endoscopist positions.

Figure 1. Axial view of the abdomen CT showing pancreatic head enhancing lesion (blue arrow) with complete left-right inverted anatomy.

Figure 2. Coronal view of the CT of the abdomen showing the dilated intrahepatic ducts (orange arrows) with complete left-right inverted anatomy.

Figure 3. Endoscopic ultrasound view showing thickened wall common bile duct and normal pancreatic duct. CBD, Common bile duct; PD, pancreatic duct.

Figure 4. Ampulla of Vater location in situs inversus totalis when ERCP is performed in the left lateral position.

Figure 5. Fluoroscopy imaging showing diffuse common bile duct stenosis. CBD, Common bile duct.

Figure 6. Fluoroscopy imaging confirming common bile duct stent placement. CBD, Common bile duct.
**TABLE 1. Summary of the reported techniques for ERCP in patients with situs inversus totalis**

| Technique | Patient position | Endoscopist position to the table (as seen from the bottom) | Key features | Advantages and challenges described |
|-----------|------------------|-------------------------------------------------------------|--------------|------------------------------------|
| Mirror image<sup>1</sup> | Right lateral | Left | Performing all regular maneuvers inversely. | Ease of cannulation of papilla. Requires manipulation with opposite hand. Changed position of room equipment. |
| 180-degree clockwise turn<sup>2,10</sup> | Prone, left lateral | Right | 180-degree clockwise rotation in stomach or duodenum. Alternatively, “pursuing endoscopy in direction inverse to usual.” | Difficulty cannulating and performing papillotomy of 1-3 o’clock papilla needing advanced papillotomy techniques. |
| Variation of limited clockwise turn<sup>15</sup> | Prone | Right | Scope inserted along the lesser curvature of the stomach, with slow clockwise rotation of the endoscope. | Ease of cannulation of central-upward ampulla. Difficulty achieving endoscopic access to duodenum. |
| 360-degree turn<sup>16-28</sup> | Prone | Right | 180-degree rotation in the stomach, then 180-degree rotation in duodenum, both in the same direction. | Difficulty controlling endoscope owing to looped shaft. Difficulty cannulating right-upward deviated ampulla. |
| Changing patient position<sup>20,21</sup> | Variable | Right | Changing position from right lateral decubitus to prone upon reaching D2; counterclockwise rotation to identify papilla. Changing position from prone to supine to locate and cannulate papilla. | Difficulty cannulating; patient intolerance. |

**DISCLOSURE**

All authors disclosed no financial relationships.

Abbreviations: CBD, common bile duct; PD, pancreatic duct; SIT, situs inversus totalis.

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