



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

Jason Naser, MD,¹ Muhammad Talal Sarmini, MD,² Catherine Vozzo, DO,² Mohannad Abou Saleh, MD,² Prabhleen Chahal, MD²

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

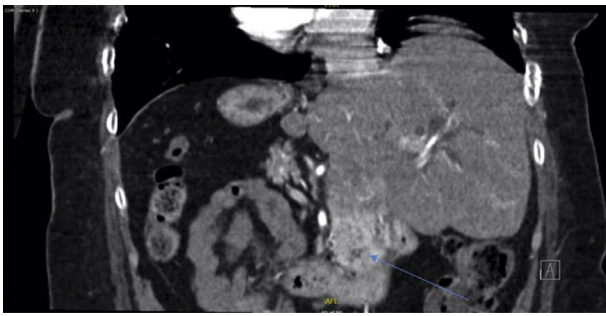


Figure 1. Axial view of the abdomen CT showing pancreatic head enhancing lesion (*blue arrow*) with complete left–right inverted anatomy.

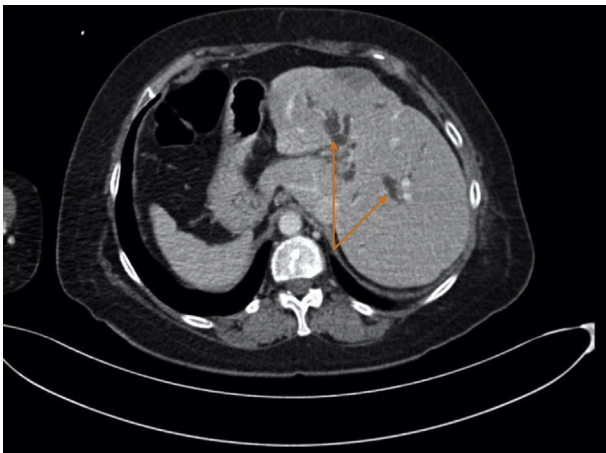


Figure 2. Coronal view of the CT of the abdomen showing the dilated intrahepatic ducts (*orange arrows*) with complete left–right inverted anatomy.



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

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.

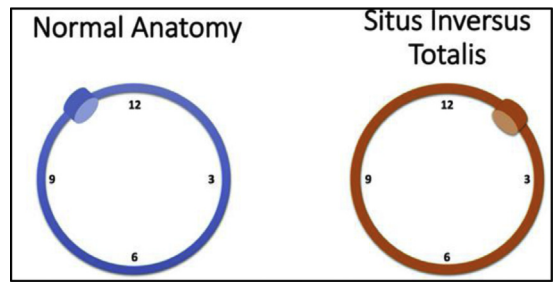


Figure 4. Ampulla of Vater location in situs inversus totalis when ERCP is performed in the left lateral position.



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

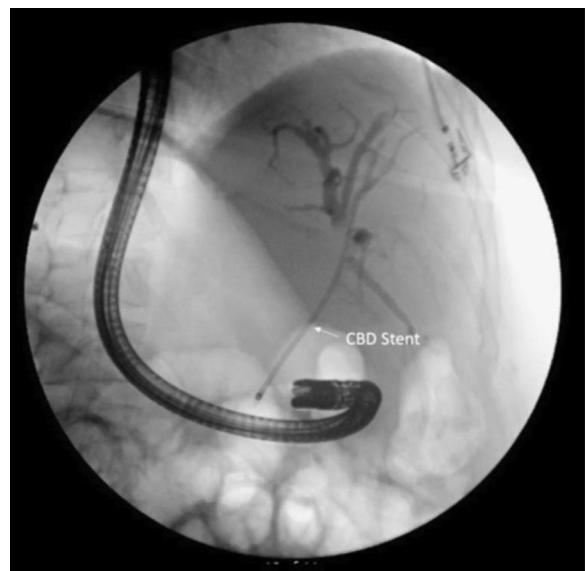


Figure 6. Fluoroscopy imaging confirming common bile duct stent placement. *CBD*, Common bile duct.

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 ³	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 ⁴⁻¹⁹	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 ²⁵	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 ²⁶⁻²⁸	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 ^{20,21}	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.

TABLE 2. Summary of the reported techniques for EUS in patients with situs inversus totalis

Patient position	Key features	Challenges described
Left lateral decubitus then right lateral decubitus ²²	Performing all regular maneuvers inversely. Linear endoscope used to perform fine-needle aspiration in conventional manner. Then switched to radial endoscope to define anatomy.	Difficulty identifying vascular anatomy. Suboptimal views of pancreas requiring changing position to right lateral decubitus following the mirror image technique to allow visualization of the entire pancreas.
Left lateral decubitus ²³	Radial endoscope used. Clockwise rotation of endoscope rather than counterclockwise to explore common bile duct. Linear scanning EUS with fine-needle aspiration.	
Left lateral decubitus ²⁴	Linear endoscope used, with insertion following inversion of usual technique.	Requiring comfortable knowledge of anatomy.

DISCLOSURE

All authors disclosed no financial relationships.

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

REFERENCES

1. Mayo CW, Rice RG. Situs inversus totalis: a statistical review of data on 76 cases with special reference to disease of the biliary tract. *Arch Surg* 1949;58:724.
2. Spoon JM. Situs inversus totalis. *Neonatal Netw* 2001;20:59-63.
3. García-Fernández FJ, Infantes JM, Torres Y, et al. ERCP in complete situs inversus viscerum using a "mirror image" technique. *Endoscopy* 2010;42:E316-7.
4. de la Serna-Higuera C, Perez-Miranda M, Flores-Cruz G, et al. Endoscopic retrograde cholangiopancreatography in situs inversus partialis. *Endoscopy* 2010;42:E98.
5. Kamani L, Kumar R, Mahmood S, et al. Therapeutic ERCP in patient with situs inversus totalis and ampullary diverticulum. *J Coll Physicians Surg Pak* 2014;24:365-6.
6. Lee JH, Kang DH, Park JH, et al. Endoscopic removal of a bile-duct stone using sphincterotomy and a large-balloon dilator in a patient with situs inversus totalis. *Gut Liver* 2010;4:110-3.
7. Sheikh I, Heard R, Tombazzi C. Technical factors related to endoscopic retrograde cholangiopancreatography in patients with situs inversus. *Gastroenterol Hepatol (N Y)* 2014;10:277-8.
8. Patel KS, Patel JN, Mathur S, et al. To twist or not to twist: a case of ERCP in situs inversus totalis. *Endoscopy* 2014;46:E304-5.
9. Morales-Rodríguez JF, Corina Cutillo E, Moreno-Loaiza O. Tratamiento quirúrgico de coledocolitiasis en situs inversus totalis: reporte de caso y revisión de la literatura [Surgical treatment of choledocholithiasis in a patient with situs inversus totalis: a case report and literature review] [in Spanish]. *Medwave* 2017;17:e7002.
10. Nordback I, Airo I. ERCP and endoscopic papillotomy in complete abdominal situs inversus. *Gastrointest Endosc* 1988;34:150.
11. Lee JM, Lee HS, Kim CD. Infundibulotomy and endoscopic retrograde cholangiopancreatography in situs inversus totalis combined with choledochoceles. *Dig Endosc* 2015;27:776.

12. Tyberg A, Lee T, Karia K, et al. Malignant obstructive jaundice in situs inversus: demonstration of precut and biliary drainage. *Gastrointest Endosc* 2015;81:1018-9.
13. Hu L, Chai Y, Yang X, et al. Duodenoscope combined with laparoscopy in treatment of biliary stones for a patient with situs inversus totalis: a case report. *Medicine (Baltimore)* 2019;98:e14272.
14. Hu Y, Zeng H, Pan X, et al. Therapeutic endoscopic retrograde cholangiopancreatography in a patient with situs inversus viscerum. *World J Gastroenterol* 2015;21:5744-8.
15. Ş Çoban, Yüksel I, Küçükazman M, et al. Successful ERCP in a patient with situs inversus. *Endoscopy* 2014;46:E222.
16. Téllez-Ávila FI, Pattel S, Duarte-Medrano G, et al. A challenging case of giant biliary stones in a patient with situs inversus totalis: conventional ERCP combined with intraductal cholangioscopy and laser lithotripsy. *Endoscopy* 2017;49:E248-9.
17. Yoshida A, Minaga K, Takeda O, et al. Successful biliary cannulation using a novel rotatable sphincterotome in a patient with situs inversus totalis. *Endoscopy* 2020;52:E333-4.
18. Shimoda F, Satoh A, Asonuma S, et al. Successful removal of multiple bile duct stones using a papillary large balloon dilation in a very elderly woman with situs inversus totalis. *Clin J Gastroenterol* 2021;14:309-13.
19. Tanisaka Y, Ryozaawa S, Sudo K, et al. Successful endoscopic retrograde cholangiopancreatography using pancreatic guidewire placement for biliary cannulation in a patient with situs inversus and Billroth-I gastrectomy (with video). *JGH Open* 2019;3:540-1.
20. Venu RP, Geenen JE, Hogan WJ, et al. ERCP and endoscopic sphincterotomy in patients with situs inversus. *Gastrointest Endosc* 1985;31:338-40.
21. Lipschutz JH, Canal DF, Hawes RH, et al. Laparoscopic cholecystectomy and ERCP with sphincterotomy in an elderly patient with situs inversus. *Am J Gastroenterol* 1992;87:218-20.
22. Kumbhari V, Abi-Hanna D, Bassan M. Gastrointestinal: endoscopic ultrasound of a pancreatic lesion in situs inversus. *J Gastroenterol Hepatol* 2014;29:229.
23. Togliani T, Pilati S, Mantovani N, et al. Extrahepatic cholangiocarcinoma in a patient with situs inversus totalis diagnosed by endoscopic ultrasound. *Endoscopy* 2013;45:E229-30.
24. Coronel M, Lanke G, Cambell D, et al. Performing endoscopic retrograde cholangiopancreatography and endoscopic ultrasound for management of malignant bile duct obstruction in a patient with a situs inversus totalis. *ACG Case Rep J* 2020;7:e00483.
25. Lee JM, Lee JM, Hyun JJ, et al. Successful access to the ampulla for endoscopic retrograde cholangiopancreatography in patients with situs inversus totalis: a case report. *BMC Surg* 2017;17:112.
26. Fiocca F, Donatelli G, Ceci V, et al. ERCP in total situs viscerum inversus. *Case Rep Gastroenterol* 2008;2:116-20.
27. Feng Q, Yao J. Hepatobiliary and pancreatic: common bile duct stones with situs inversus totalis. *J Gastroenterol Hepatol* 2020;35:12.
28. El Hajj II, Sherman S, Ceppa EP, et al. ERCP and laparoscopic cholecystectomy in a patient with situs inversus totalis. *Dig Liver Dis* 2017;49:1374.

Department of Internal Medicine, Cleveland Clinic, Cleveland, Ohio (1), Department of Gastroenterology and Hepatology, Digestive Disease Institute, Cleveland Clinic, Cleveland, Ohio (2).

Copyright © 2022 American Society for Gastrointestinal Endoscopy. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

<https://doi.org/10.1016/j.vgie.2022.05.008>

Facebook

Follow *VideoGIE* on Facebook to receive the latest news, updates, and article links. Visit <https://www.facebook.com/videogiejournal/> and keep checking back to view our most recent posts.