

CASE REPORT

Successful Endoscopic Retrograde Cholangiopancreatography for Management of Choledocholithiasis in a Patient With Situs Inversus Totalis: A Case Report and Literature Review



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Situs inversus totalis is a rare congenital disorder defined by the transposition of all viscera to the opposite side of the body. Because of this anatomical alteration, endoscopic retrograde cholangiopancreatography (ERCP) in such a population is significantly challenging. Herein we report a case of a 50-year-old woman presented with epigastric and left upper quadrant pain. Preoperative examination revealed abnormal liver chemistries, situs inversus totalis, and multiple distal common bile duct stones with intrahepatic and extrahepatic bile duct dilatation on abdominal ultrasound. The patient underwent a successful ERCP to relieve biliary obstruction. It was performed while the patient was placed in a supine position with the endoscopist standing on the left side of the patient. The major emphasis in ERCP is adjusting the patient and endoscopist positions during the procedure. The advantage of the supine position is that the endoscope could easily reach the pyloric ring and, after reaching the duodenum, the papilla could be reached without difficulty.

Keywords: Situs inversus totalis (SIT); Endoscopic retrograde cholangiopancreatography (ERCP); Choledocholithiasis; CBD stones; Biliary cannulation

Herein, we report a case of SIT who underwent ERCP for the management of choledocholithiasis. We also include a review of the literature of related reports. To our knowledge, this case is the first reported in Saudi Arabia.

Case Report

A 50-year-old female presented to the emergency department with acute onset of epigastric and left upper quadrant pain. She is known to have situs inversus and had undergone cholecystectomy for cholelithiasis more than 10 years before. The operation was started as laparoscopic and then was converted to open due to anatomical anomalies. At the time of presentation, she was in severe pain, but had normal vital signs, with mild tenderness on palpation in the epigastric area. Other aspects of her clinical exam were unremarkable other than what is consistent with her dextrocardia.

Her laboratory investigations are as follows: white cell count: 11.8; serum amylase: 1694 U/L (normal less than 125); total bilirubin: 70.8 umol/L (normal less than 20.50, direct bilirubin: 57.6, alanine transaminase: 482, aspartate aminotransferase: 411); and alkaline phosphatase: 204. Plain chest X-Ray (Figure 1) was normal except for the finding of dextrocardia. However, there were no cardiovascular physiological disturbances that affected her case. An abdominal ultrasound (Figure 2) was remarkable for finding the liver in the left upper quadrant, and mild intrahepatic and extrahepatic bile duct dilatation with multiple distal common bile duct (CBD). The patient then underwent an ERCP to relieve biliary obstruction due to the presence of choledocholithiasis. It was performed under general anesthesia with endotracheal intubation in a supine

Introduction

Situs inversus totalis (SIT) is a rare autosomal recessive genetic condition, which is characterized by a left-right transposition of all viscera as a mirror image.¹ It occurs at a frequency of 1 in 6000 to 1 in 80,000 live births.²

This uncommon anatomy requires special attention with invasive interventions, especially endoscopic treatment. Studies conducted on this issue are less than 50 in PubMed to date because of its rarity. They were case reports which present variation on endoscope manipulation and patient-endoscopist position.^{3,4} However, the optimal technique and patient-endoscopist position during endoscopic retrograde cholangiopancreatography (ERCP) in SIT patients are still unclear.



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2772-5723

<https://doi.org/10.1016/j.gastha.2024.09.010>

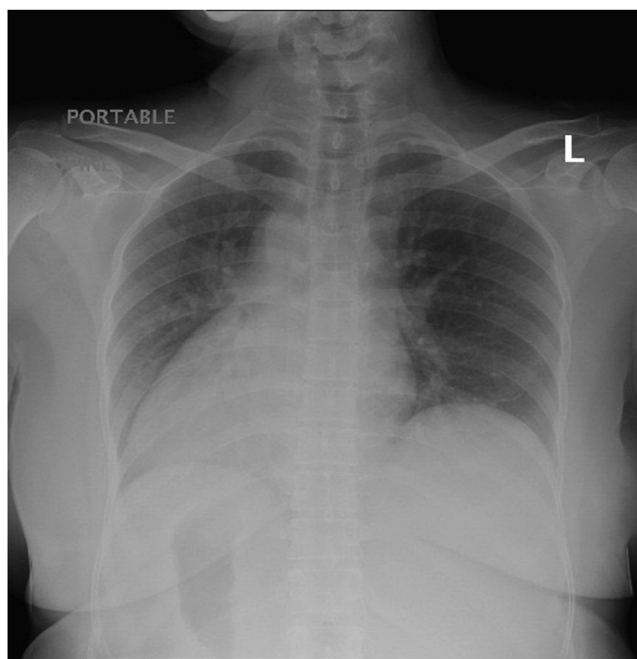


Figure 1. Chest X-ray revealed dextrocardia.

position with the endoscopist standing on the left side of the patient.

The duodenoscope was introduced into the stomach; it was easy to intubate the duodenum. Once into the duodenum, the scope was advanced further and rotated anticlockwise with tip deflection, leading to adequate visualization of the duodenal papilla, and easy access and deep cannulation of the CBD. The papilla was very prominent with the suggestion of an impacted stone (Figure 3B). The orifice was oriented caudally making it difficult to gain deep cannulation of the CBD. The prominent papilla, however, made it relatively easy to perform a precut needle-knife and gain deep access into the CBD. The needle-knife was then exchanged to a standard wire-guided sphincterotome, and the sphincterotomy was extended. Diluted contrast was injected revealing large rhomboid shape filling defects in the CBD, with mild proximal ductal dilatation. The filling defects were proven to be stones when extracted using a balloon extraction catheter. A balloon sweep of the bile duct confirmed complete clearance of the bile duct from any filling defect. There was no immediate or delayed complication from the procedure, such as bleeding, pancreatitis, or perforation. The patient did well post-operatively with remarkable improvement in the liver tests. She was discharged home 2 days postprocedure.

Discussion

ERCP is the standard procedure for the removal of CBD stones.⁵ It is usually performed with patients of normal anatomical structure in a left prone position or a modified left-lateral position.^{6,7} These positions are clearly superior to the supine position in terms of difficulty of cannulation,

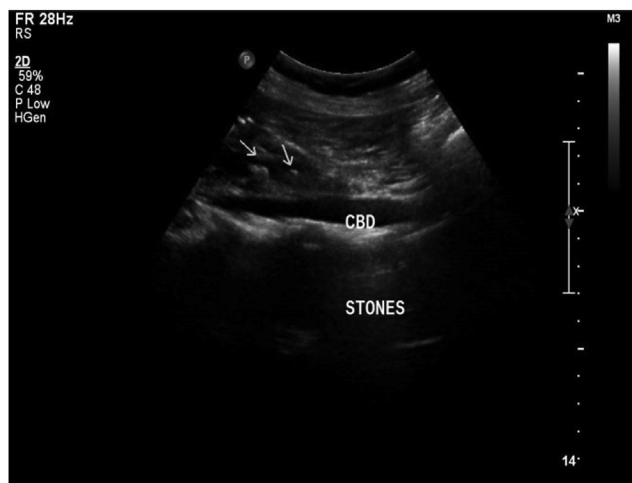


Figure 2. Intrahepatic and extrahepatic bile duct dilatation with multiple distal CBD stones.

safety, and possibly tolerability. However, in some situations, a supine position is advocated. For example, for patients who require deep sedation with endotracheal anesthesia, or have cardiopulmonary dysfunction, extreme obesity, massive ascites, and advanced pregnancy, the prone position is difficult to adopt.⁸

Concerning patient positioning, in most reported cases of SIT, the patient was placed in the prone position as detailed in Table A1. The supine position was also sporadically reported.^{1,9-11} Previous reports of ERCP procedures done in patients with SIT are summarized in Table A1.

When the patient with normal anatomy is placed in a prone position, the disposition of the organs is almost the same as that of a patient with a situs inversus in the supine position. Thus, Rocha et al. found that the supine position makes the procedure easier and more effective, like the procedure performed in patients without situs inversus.⁹

Our case is the fourth reported case where a patient with underlying SIT was maintained in a supine position throughout the procedure.

ERCP procedures are challenging in the presence of anatomical abnormalities. The currently available literature on ERCPs performed in patients with SIT has been restricted to case reports due to its rarity. There are many technical difficulties in patients with SIT who underwent ERCP such as patient-endoscopist position, access to duodenal papilla, and maneuverability of scope.¹² Cannulation is significantly affected in SIT patients due to the following reasons: 1) The direction of biliary orifice is inverted to 1–3 o'clock instead of 10 to 11 o'clock in the normal population, therefore selective biliary cannulation maneuvers should be performed inversely as per normal procedures. 2) Scope shortening in the duodenum may be more difficult when the scope had to be rotated through 180°.⁴

To improve the success rate, various techniques to access the ampulla have been described in patients with situs inversus.¹³ In one technique, the endoscope is rotated 180°

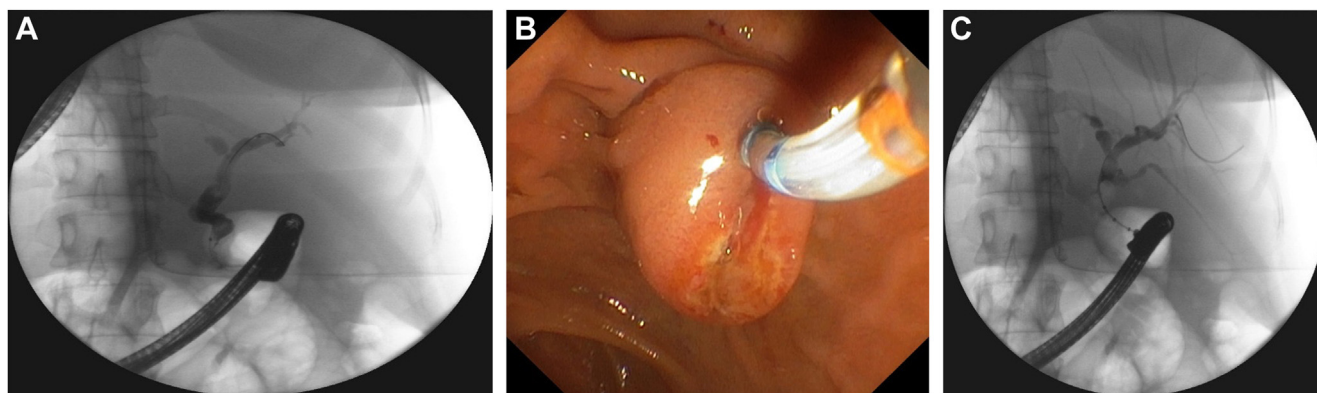


Figure 3. (A) A cholangiogram reveals multiple CBD filling defects. Note the long position of the duodenoscope. (B) The prominent papilla which suggested impacted stone. (C) Occlusion cholangiogram relieving clearance of the duct.

clockwise in the stomach; and again while in the second part of the duodenum.¹⁴ Rotatable sphincterotome is proved to be useful in several cases.^{15–17} Another option is a “mirror image” method. The patient is usually placed in the right prone position to manage the reversal of internal organs, so all endoscopic maneuvers are performed inversely.¹⁸

As illustrated in Table A1 in patients with situs inversus, endoscopists have variable approaches to the performance of ERCP. Most seem to favor the “mirror-image” technique, in a prone position, with the endoscopist being on the right side of the patient. In our case, the patient was placed in the supine position, and the endoscopist maintained the usual position; being on his left side. The advantage of the supine position is that the endoscope could easily reach the pyloric ring, and after reaching the duodenum, the papilla could be reached without difficulty by the pushing method, together with rotation and the usual stretching method.¹⁹

Complications of ERCP in SIT patients were rare according to published cases. Lakhtakia S et al. reported that 1 case of bleeding from portal biliopathy during stone removal was successfully treated by self-expandable metal stent and balloon compression.²⁰ In previous case series have shown that complication of ERCP occurred in 3 patients (3 of 14, 21.4%), including 1 bleeding, 1 pneumonia and 1 acute myocardial infarction. Compared to patients in modified position, those in prone position had numerically less successful cannulations (66.7% vs 90.9%) and higher adverse events (33.3% vs 18.2%).²¹

Overall, the key to success in such cases is the understanding of the abnormal anatomy and performing the procedure by an expert trying various maneuvers.

We report a case of SIT presenting with choledocholithiasis, in whom ERCP was successfully performed in the supine position. Till now, no standard position for ERCP in SIT, but an appropriate position can be selected based on the patient's condition and the practice ie endoscopist is more familiar with. ERCP is generally feasible and safe in patients with situs inversus. To ensure higher success rates of procedure and minimize the incidence of complications, it is best to be operated by skilled

endoscopists. Current available literature in this subject has been restricted to case reports due to its rarity. Therefore, more prospective studies and larger cohorts are required to determine the optimal approach as well as to further characterize the experiences and outcomes of ERCP in patients with situs inversus.

Supplementary Materials

Material associated with this article can be found in the online version at <https://doi.org/10.1016/j.gastha.2024.09.010>.

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Received October 21, 2023. Accepted September 16, 2024.

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Conflicts of Interest:

The authors disclose no conflicts.

Funding:

The authors report no funding.

Ethical Statement:

The corresponding author certifies on behalf of all coauthors that the institutional has approved the protocol for investigations involving human or animal subjects. Furthermore, all related research has been conducted in accordance with the ethical and humane principles of research.

Reporting Guidelines:

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