

# Outcomes of endoscopic retrograde cholangiopancreatography in patients with situs inversus viscerum

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Patient #	Indication	Clinical Success	Technical Success	Reason for failure	Rendezvous ERCP attempted?	Rendezvous ERCP successful?	Adverse events
1	Choledocholithiasis	Yes	Yes				No
2	Pancreatic adenocarcinoma	Yes	Yes				No
3	Choledocholithiasis with cholangitis	No	No	Unable to cannulate the ampulla	Yes	Yes	No
4	Periampullary adenocarcinoma	Yes	Yes				No
5	Choledocholithiasis	No	No	Unable to cannulate the ampulla	Yes	Yes	No
6	Choledocholithiasis	Yes	Yes				No
7	Benign biliary stricture	Yes	Yes				No
8	Choledocholithiasis with cholangitis	No	No	Unable to cannulate the ampulla	Yes	Yes	No

**In patients with sinus inversus viscerum, ERCP with interventional radiology-guided rendezvous appears both safe and efficacious in cases in which biliary cannulation failed during the index ERCP.**

Clin Endosc 2023; 56: 790-794

Received: November 3, 2022    Revised: January 4, 2023  
 Accepted: January 20, 2023

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**Background/Aims:** Situs inversus viscerum (SIV) is a congenital condition defined by left-to-right transposition of all visceral organs. This anatomical variant has caused technical challenges in endoscopic retrograde cholangiopancreatography (ERCP). Data on ERCP in patients with SIV are limited to case reports of unknown clinical and technical success rates. This study aimed to evaluate the clinical and technical success rates of ERCP in patients with SIV.

**Methods:** Data from patients with SIV who underwent ERCP were retrospectively reviewed. The data were collected by querying the nationwide Veterans Affairs Health System database for patients diagnosed with SIV who underwent ERCP. Patient demographics and procedural characteristics were collected.

**Results:** Eight patients with SIV who underwent ERCP were included. Choledocholithiasis was the most common indication for ERCP (62.5%). The technical success rate was 63%. Subsequent ERCP with interventional radiology-assisted rendezvous has increased the technical success rate to 100%. Clinical success was achieved in 63% of cases. Among cases of subsequent rendezvous ERCP after conventional ERCP failure, clinical success was achieved in 100%.

**Conclusions:** The clinical and technical success rates of ERCP in patients with SIV were both 63%. In patients with SIV in whom ERCP fails, interventional radiology-assisted rendezvous ERCP can be considered.

**Keywords:** Endoscopic retrograde cholangiopancreatography; Situs inversus

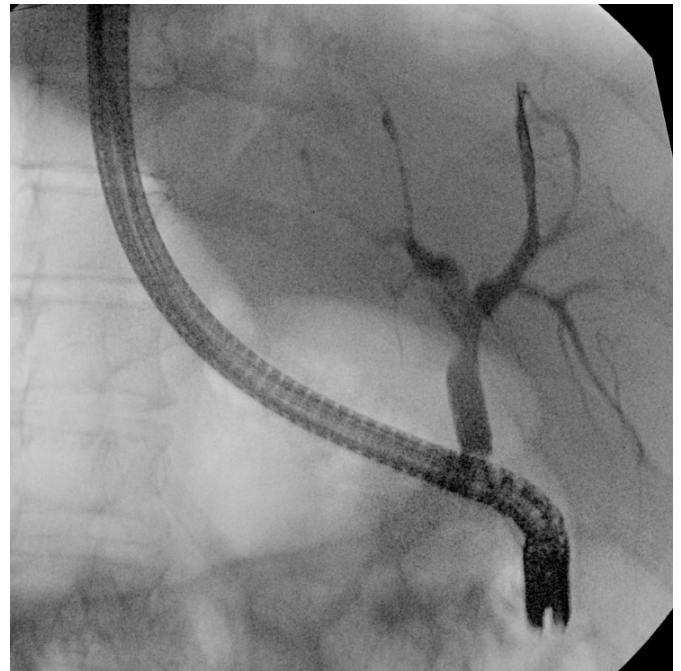
## INTRODUCTION

Situs inversus viscerum (SIV) is a rare congenital condition with a prevalence of 1 of every 10,000 individuals.<sup>1</sup> This condition is defined by the complete left-to-right transposition of all major visceral organs, including the heart and liver, from their normal positions (Fig. 1).<sup>2</sup> Anatomical abnormalities create technical challenges during endoscopic and surgical procedures.<sup>3,4</sup>

Endoscopic retrograde cholangiopancreatography (ERCP) is an advanced endoscopic procedure during which a side-viewing endoscope is advanced into the duodenum, allowing for various instruments to be passed through the ampulla of Vater into the biliary and pancreatic ducts for managing various pancreaticobiliary pathologies. ERCP-guided interventions are commonly used to manage pancreaticobiliary conditions, including choledocholithiasis or biliary obstruction. The success rate of ERCP for the management of pancreaticobiliary disease in patients with normal anatomy reportedly exceeds 95%.<sup>5,6</sup> The success rate of ERCP in patients with SIV is unknown and data are limited to case reports.<sup>7</sup> This study aimed to report the outcomes of therapeutic ERCP in patients with SIV using data collected from a large national database.

## METHODS

A retrospective chart review of the nationwide Veterans Health Administration Services electronic database was performed for patients diagnosed with SIV (International Classification of Diseases [ICD] ninth revision code: 759.3; ICD tenth revision



**Fig. 1.** Fluoroscopy image demonstrating a mirror image of cholangiogram during endoscopic retrograde cholangiopancreatography for patient with situs inversus viscerum.

code: Q89.3) between January 1, 2010, and March 31, 2022; who underwent ERCP (current procedural terminology: 43260-43265, 43277, 43278) for management of pancreaticobiliary disease. The demographic, clinical, radiologic, and endoscopic data were collected in our study. The demographic information collected included age, sex, and ethnicity. The clinical data collected included indications for ERCP, procedural outcome

(clinical success), and post-procedural adverse events, such as bleeding, infection, perforation, and post-ERCP pancreatitis. Endoscopic data collected included procedural documentation, such as body position and maneuver(s) (if attempted) to cannulate the ampulla and technical success. Follow-up endoscopic data were also collected, such as repeat ERCP with rendezvous attempts, technical success, and overall adverse events (pancreatitis, bleeding, and perforation). Technical success was defined as successful biliary cannulation, while clinical success was defined as stone removal or successful biliary drainage. Statistical analyses were performed using Microsoft Excel (ver. 2206; Microsoft 365).

**Ethical statements**

The study was conducted according to the guidelines of the Declaration of Helsinki, and approved by the Institutional Review Board of Minneapolis Veteran Affairs Health System (IRB# 1648481 and date of approval May, 10 2022).

**RESULTS**

In total, 654 patients diagnosed with SIV were identified in the database. Eight patients underwent ERCP for the management of various pancreaticobiliary diseases. The most common indication was choledocholithiasis (63%), followed by malignancy (25%) and benign biliary stricture (12%). Most patients were men and self-identified as White. The median age was 68 years.

The technical and clinical success rates of index ERCP were both 63%. The inability to cannulate (25%) or visualize (13%) the ampulla despite positional changes was cited as the cause of technical failure during the index ERCP.

All patients in whom the index ERCP failed underwent a successful follow-up ERCP with interventional radiology (IR)-assisted rendezvous. The overall success rate of ERCP for managing biliary obstruction in patients with SIV was 100%, including cases of repeat ERCP with IR-assisted rendezvous. No clinical or technical adverse event were reported in our cohort (Table 1).

**DISCUSSION**

The success rate of ERCP for managing pancreaticobiliary disease reportedly exceeds 95%.<sup>8</sup> Currently, data are lacking on the efficacy of ERCP for managing pancreaticobiliary disease in patients with SIV. The success rate of the first-time ERCP in the current population was much lower (63% vs. >95%). This relatively low success rate is likely due to the altered anatomy of SIV patients. Despite the short path to the ampulla, conventional movements with a side-viewing scope that allows for reaching the ampulla in normal anatomy are not always possible; therefore, visualizing the ampulla can be challenging. Furthermore, the movements required to achieve and maintain a stable endoscope position in the duodenum differ, which can make selective biliary cannulation challenging.

**Table 1.** Patient demographic variables, procedural characteristics, and adverse events

Sex	Race	Age (yr)	Indication	Clinical success	Technical success	Reason for failure	Rendezvous ERCP attempted?	Rendezvous ERCP successful?	Adverse events
Male	White	72	Choledocholithiasis	Yes	Yes	-	-	-	No
Male	African American	74	Pancreatic adenocarcinoma	Yes	Yes	-	-	-	No
Male	White	88	Choledocholithiasis with cholangitis	No	No	Unable to cannulate the ampulla	Yes	Yes	No
Male	White	62	Periampullary adenocarcinoma	Yes	Yes	-	-	-	No
Male	White	59	Choledocholithiasis	No	No	Unable to cannulate the ampulla	Yes	Yes	No
Male	White	88	Choledocholithiasis	Yes	Yes	-	-	-	No
Female	Native American	63	Benign biliary stricture	Yes	Yes	-	-	-	No
Male	Hispanic	63	Choledocholithiasis with cholangitis	No	No	Unable to visualize the ampulla	Yes	Yes	No

ERCP, endoscopic retrograde cholangiopancreatography; -, not applicable.

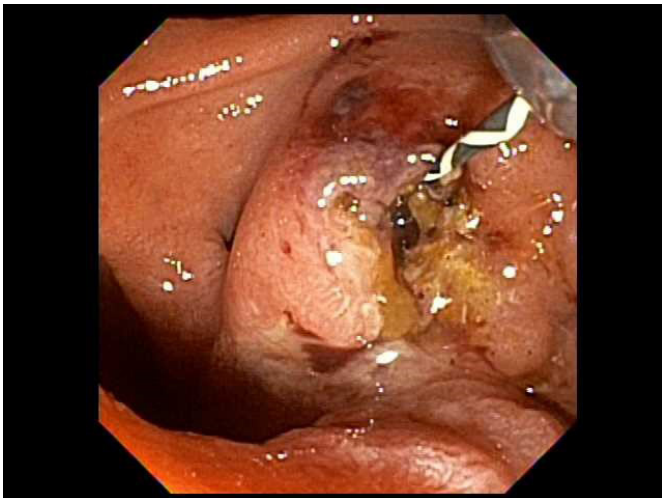
Previous case reports suggested that several maneuvers can increase technical success rates. One approach is the mirror image technique, in which the patient is placed in the right lateral decubitus position with the radiosurgical equipment placed at the back. All endoscopic maneuvers were performed inversely as per normal procedures, as if the procedure was a mirror reflection of the standard procedure.<sup>9</sup> Another approach is to rotate the duodenoscope by 180° clockwise in the stomach or duodenum. However, this approach may present difficulty in cannulating the papilla, which is usually in the 1 to 2 o'clock position as opposed to the 11 o'clock position in patients without SIV (Fig. 2).<sup>10-12</sup> Other case reports suggested changing the patient's position from right lateral decubitus to prone upon reaching the second duodenal portion to facilitate papillary identification and cannulation.<sup>7</sup> Other experts suggested rotating the duodenoscope counterclockwise while in the duodenum with the left arm hanging straight down while advancing from the duodenal bulb to the second duodenal portion.

ERCP with IR-assisted rendezvous is a combined technique in which an intrahepatic biliary tree is accessed through the liver and a guidewire is advanced percutaneously from the intrahepatic biliary tree across the major duodenal ampulla to facilitate bile duct cannulation during ERCP (Fig. 3). This technique has been utilized in different clinical scenarios as well as in patients with SIV, but existing data are limited to case reports.<sup>7,13-16</sup> Guidewire placement allows for better ampulla vi-

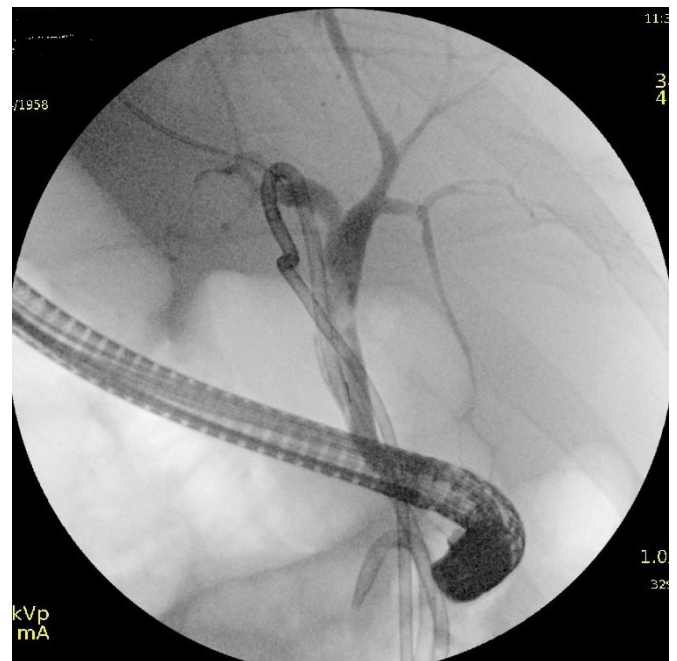
sualization and/or allows endoscopic biliary cannulation using a catheter over a guidewire or another guidewire adjacent to the percutaneously placed guidewire. In one case, the patient had multiple duodenal diverticula and the ampulla was not visible in the index ERCP despite extensive interrogation and multiple positional changes. In this case, the ampulla was located within the periampullary diverticulum. Therefore, after internal-external drain placement by IR, a rendezvous procedure was performed and endoscopic biliary cannulation was achieved by following the guidewire into the bile duct (Table 1). This maneuver increased the overall technical and clinical success rates of ERCP in our cohort to 100%. In addition, we did not identify any clinically significant adverse events associated with this procedure.

In our limited experience, repeat ERCP with IR-guided rendezvous appears both safe and efficacious in cases in which biliary cannulation failed during the index procedure. It should be considered the next step after standard ERCP failure (Fig. 3) at centers featuring the required expertise.

Our study had several limitations. First, it included a small sample size; however, given the rarity of SIV patients who develop pancreaticobiliary disease, it would be difficult to collect a sufficiently large sample size. Second, none of the patients in



**Fig. 2.** Endoscopic view of biliary sphincterotomy in a patient with situs inversus viscerum with direction sphincterotomy toward the 1 o'clock position (versus 11 o'clock position in patients with a normal anatomy).



**Fig. 3.** Fluoroscopy cholangiogram of patient with situs inversus viscerum and presence of percutaneous biliary drain at time of interventional radiology-assisted rendezvous procedure.

whom biliary cannulation failed on the index ERCP had undergone an endoscopic ultrasound (EUS)-guided rendezvous attempt; therefore, it is unclear whether EUS-guided rendezvous is feasible in this cohort. Despite these limitations, our study is the largest case series to report the ERCP outcomes of patients with SIV. Despite the complete transposition of all major visceral organs, the index ERCP was safe and had an acceptable technical/clinical success rate. Our study findings suggest that ERCP should be attempted prior to the percutaneous approach in patients with SIV requiring biliary intervention.

### Conflicts of Interest

The authors have no potential conflicts of interest.

### Funding

None.

### Author Contributions

Conceptualization: LL, DS; Data curation: LL, NM, AW, DS; Formal analysis: LL; Methodology: LL, DS; Software: AW; Supervision: MB, DS; Writing—original draft: LL, DS; Writing—review & editing: all authors.

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