



## Case report

## Successful endoscopic drainage of malignant obstructive jaundice in patients with situs inversus totalis: Two cases report

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## ARTICLE INFO

## Keywords:

Situs inversus totalis  
Endoscopic retrograde cholangiopancreatography  
Percutaneous transhepatic cholangial drainage  
Gallbladder carcinoma  
Jaundice

## ABSTRACT

**Introduction and importance:** Although endoscopic techniques in situs inversus totalis (SIT) have been reported, endoscopic retrograde cholangiopancreatography (ERCP) in patients with situs inversus totalis (SIT) remains difficult to every hepatobiliary surgeon. To investigate the differences of each position, ERCP was used to perform through two different body positions.

**Case presentation:** Herein we report a 63-year-old woman presented with epigastric pain for 2 months and jaundice for 7 days and a 51-year-old man with presented jaundice for 7 days. Preoperative examination revealed situs inversus totalis and gallbladder carcinoma with diffuse dilatation of the biliary tree. ERCP was used to perform by using two different body positions. In addition, the ERCP combined with percutaneous transhepatic cholangial drainage (PTCD) was performed in the second patient.

**Clinical discussion:** The different endoscopic approaches are used in different positions, the endoscopist should be familiar with mirror symmetrical anatomy. We argue that the prone position has a higher surgical success rate and ERCP combined with PTCD will be easier in SIT patients.

**Conclusion:** ERCP in SIT patients is generally safe and it will be easier by combining with PTCD.

## 1. Introduction

Situs inversus is a very rare congenital malformation in which the anatomy of the left and right sides is reversed, like in a mirror image from their normal positions. The incidence of this generally autosomal congenital recessive genetic condition is approximately 1 in 6000 to 1 in 80,000 [1]. Although the patients have mirrored internal organs, there is no affect in their daily life with completely normal viscera function. When they have medical symptoms, it will bring great difficulties to clinical diagnosis and therapy, especially endoscopic treatment [2].

Herein, we report two cases of situs inversus totalis (SIT) who underwent ERCP for the gallbladder carcinoma by using two different positions and compare the merits and demerits of each position. The study was approved by the patients. This work has been reported in line with the SCARE criteria [3].

## 2. Case reports

## 2.1. Case 1

A 63-year-old woman presented to our hospital in 2019 with a dull pain in the upper abdomen for 2 months and jaundice for 7 days. Laboratory studies showed that the alanine transaminase was 392 U/L, the aspartate transaminase was 263 U/L, total bilirubin was 11.16 mg/dL and direct bilirubin was 6.00 mg/dL. The alkaline phosphatase level was 1049 IU/L and gammaglutamyl transpeptidase was 1152 U/dL. The carcinoembryonic antigen was 14.01 µg/L. All other laboratory parameters (e.g., Carbohydrate antigen 19-9) were in the normal range. A chest X-ray (Fig. 1) confirmed dextrocardia. Computed tomography (CT) demonstrated gallbladder carcinoma and total SIT in the abdomen (Fig. 2A). The diffuse dilatation of biliary tree and multiple intrahepatic metastases were also found on CT (Fig. 2B).

After full preoperative preparation, ERCP was performed with a side-viewing endoscope (Olympus TJF-260). The patient was in a left prone position, which is a common position for ERCP with the endoscopist standing at the side of patient right hand. The endoscope is easy to loop

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Received 21 January 2022; Received in revised form 19 February 2022; Accepted 22 February 2022

Available online 25 February 2022

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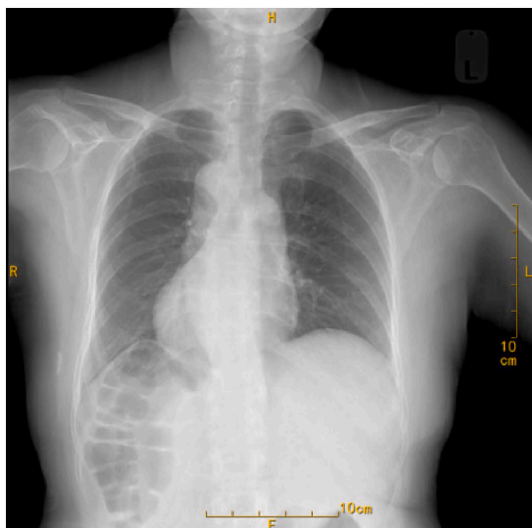


Fig. 1. Chest X-ray revealed dextrocardia.

in the fundus of the stomach after entering the gastric cavity (Fig. 3A), so the endoscopic approach was contrary to the normal. Then the endoscope was rotated 180° clockwise to the lower right along the lining of the stomach until keeping the pylorus in setting-sun position (Fig. 3B). The endoscope was turned by moving the hand counterclockwise and rotating the up-down button clockwise with turning the endoscope tip down before the duodenal papilla was seen. Putting the duodenal papilla in the front by rotating the right-left button of the endoscope to the left and straightening the endoscope, then used the cutting knife with pre-loaded guide wire to try intubation toward the direction of 1 o'clock of the opening of the papilla (Fig. 3C). Cholangiography showed that the biliary ducts had diffuse dilatation (Fig. 3D). After a standard sphincterotomy (Fig. 3E), an expandable metallic biliary endoprosthesis (EMBE) was placed (Fig. 3F), at the same time X-ray revealed there was no looped endoscope shaft (Fig. 3G). A postoperative abdomen X-ray showed that the metal stent was fixed in place (Fig. 3H). The trick to success depended on keeping remember that the anatomy of SIT was mirrored from left to right, not from up to down. Therefore, when the endoscopist performed endoscope up and down operations, which was the same as their usual operating habits in a normal person, and when performed endoscope left and right operations, which was opposite to their usual operating habits in a normal person.

## 2.2. Case 2

A 51-year-old male was admitted to the local hospital because of

jaundice for 7 days. He was diagnosed as gallbladder carcinoma with SIT and underwent expectant treatment. He visited our hospital due to surgical treatment. Laboratory examination showed that the aspartate transaminase was 70 U/L, total bilirubin was 15.60 mg/dL and direct bilirubin was 9.25 mg/dL. The alkaline phosphatase level was 342 IU/L and  $\gamma$ -glutamyltransferase level was 2624 U/dL. The Carbohydrate antigen 19-9 was 58.41 U/mL. Chest X-ray (Fig. 4) demonstrated SIT. Computed tomography (CT) and magnetic resonance cholangiopancreatography (MRCP) revealed gallbladder carcinoma had invaded the bile ducts on both sides and diffuse dilatation of biliary tree (Figs. 5 and 6).

In order to relieve jaundice, the patient underwent percutaneous transhepatic cholangial drainage (PTCD) first. Three days later, ERCP was performed in a supine position because the patient wanted to have stomach examination at the same time with the endoscopist standing at the side of patient left hand. The endoscope was rotated 180° counterclockwise passing through the stomach and duodenum just as normal people in the left prone position due to the mirror anatomy, therefore we could easily see the duodenal papilla. After repeatedly failed intubation toward the direction of 11 o'clock of the opening of the papilla, injecting the contrast medium in PTCD showed the bile duct (Fig. 7A). The duodenal papilla was punctured by a guide wire through PTCD tube (Fig. 7B) and docked with a cutting knife under endoscope (Fig. 7C). When the guide wire inside the cutting knife was passed up into the bile duct (Fig. 7D), an expandable metallic biliary endoprosthesis (EMBE) was placed (Fig. 7E). An intraoperative X-ray showed that the metal stent was fixed in place (Fig. 7F).

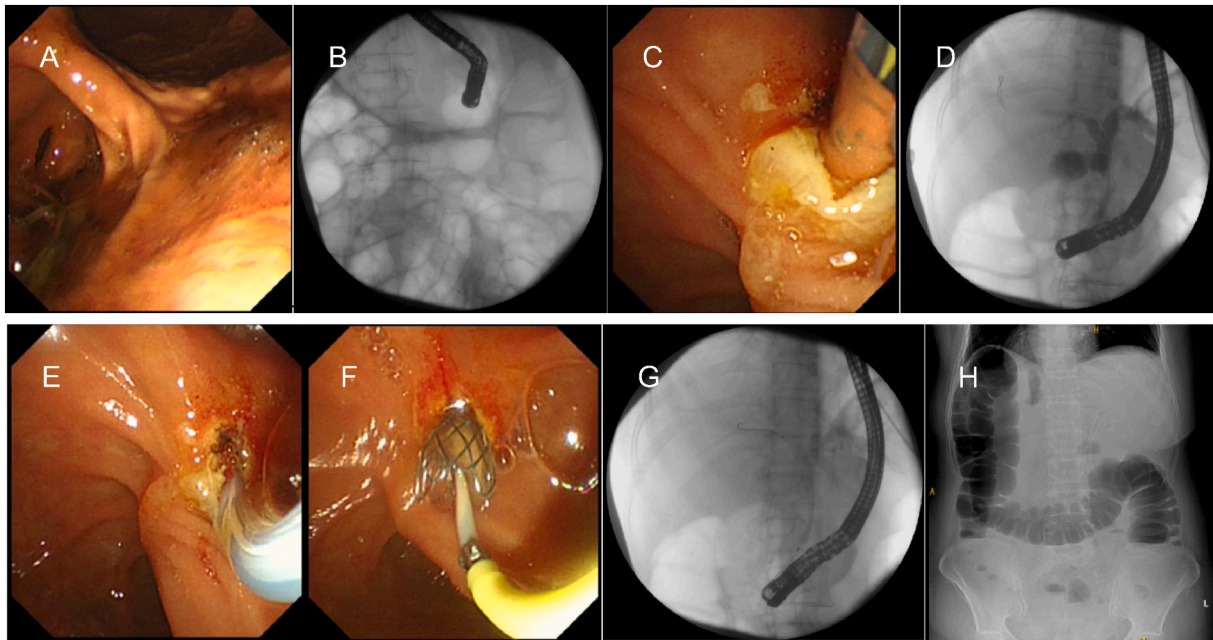
Postoperative laboratory results showed that their serum total bilirubin and direct bilirubin levels were within the normal ranges, postoperative complications such as pancreatitis and cholangitis did not occur. They were discharged in good condition. Both patients were unable to undergo radical surgery because of the female patient whose TNM was T3N<sub>x</sub>M0 had multiple liver metastases and the male patient whose TNM was also T3N<sub>x</sub>M0 had invaded both left and right intrahepatic bile ducts. Unfortunately, the female patient died 6 months after biliary drainage with refusing chemotherapy, and the male patient died after 11 months with multiple chemotherapy. But the life quality of both patients had improved during this time.

## 3. Discussion

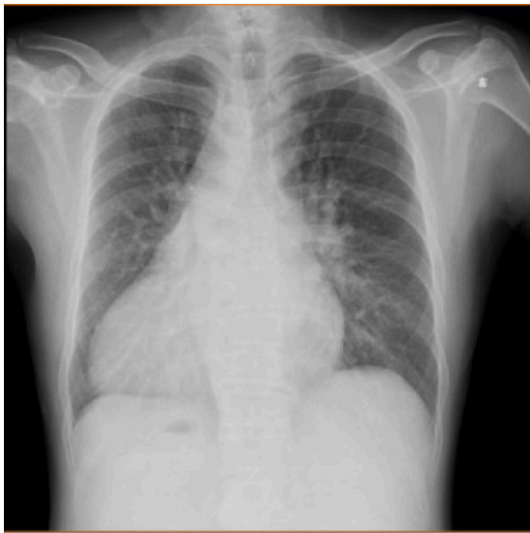
ERCP has been widely used in the minimally invasive diagnosis and treatment of various biliary and pancreatic diseases due to its advantages of less trauma and quick postoperative recovery. However, congenital anatomical abnormalities change the normal way of inserting the endoscope and increase the difficulty of selective biliopancreatic duct intubation, which makes ERCP more challenging. When a common bile duct cannot be selectively cannulated in patients with malignant obstructive jaundice, the PTCD-assisted over-the-wire cannulation



Fig. 2. A CT revealed gallbladder carcinoma and total SIT in the abdomen. B CT revealed diffuse dilatation of biliary tree and multiple intrahepatic metastases.



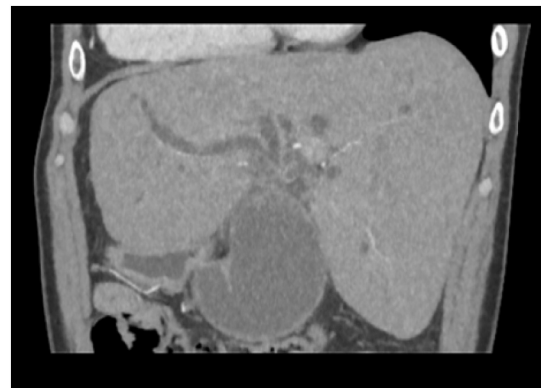
**Fig. 3.** A Entered the gastric cavity. B Rotated the endoscope 180° clockwise in the stomach. C Intubated the papilla at 1 o'clock. D Showed that the biliary ducts by Cholangiography. E Made an endoscopic papillotomy toward the direction of 1 o'clock. F Released the EMBE under endoscope. G Released the EMBE under X-ray. H Reviewed the EMBE was fixed in place.



**Fig. 4.** Chest X-ray demonstrated SIT.

method may be available [4].

Situs inversus viscerum (SIV) is a congenital anatomical abnormality, which may be related to chromosomal structural aberration, chromosomal number change, rotation disorder during fetal development and family inheritance [5]. According to the organs involved, it includes situs inversus partialis (SIP) and situs inversus totalis (SIT), the latter is more common in clinical practice. ERCP is generally performed in a left prone position with patients of normal anatomical structure. To date, the majority of SIT patients treated with ERCP used in the prone position and the left lateral position [6–9]. The supine position and the right lateral position were also sporadically reported [10,11]. In 1988, successful ERCP in patients with SIT was reported by using a prone position and Nordback et al. [12] found that the common bile duct was selectively cannulated toward the direction of 1 o'clock. Since then, some



**Fig. 5.** CT revealed diffuse dilatation of biliary tree.

authors have reported SIT cases treated with ERCP in other positions. Chowdhury et al. [13] reported a case in which patient was placed in the left lateral position with the endoscopist standing the left side of the table in 1997. The duodenal papilla was put in the front by rotating a 90° twist to the left and straightening the endoscope. Subsequently, the intubation of selectively common bile duct was performed successfully. Case with the endoscopist at the right side of the table was also reported by using a prone position. Fiocca et al. [14] reported that the endoscope was rotated 180° clockwise in the stomach and the duodenum. A similar case was turning the endoscope 180° clockwise in the stomach and using a rotating sphincterotome for intubation [15]. However, these cases were performed in a usual environment such as the position of the patient, the endoscopist and the endoscopic machine. It is difficult to adjust and fix the position of the duodenal papilla, the intubation and cholangiography are not easy to succeed. Therefore, a skilled endoscopist with extensive experience is extremely important. Hu Y et al. [2] reported a case in which the patient was initially in a supine position with the endoscopist standing at the left side of the patient who faced to the left, then the patient was adjusted to a right prone position due to



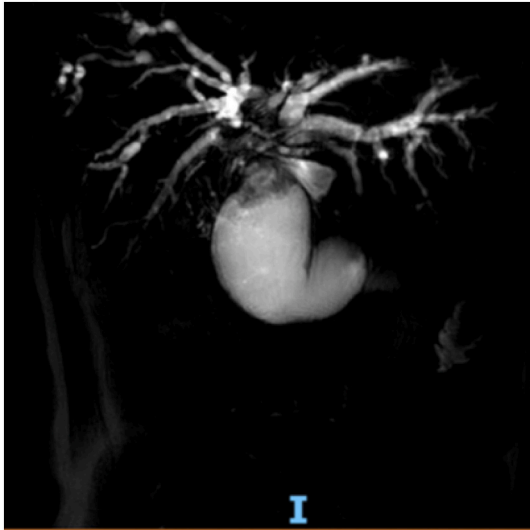


Fig. 6. MRCP revealed gallbladder carcinoma.

great difficulty in intubation. The method is exactly that Fernández et al. [11] reported a “mirror image” technique, but which is inconvenient shortening the endoscope by rotating counterclockwise, so the endoscopist required to operate the endoscope with the right hand. In addition, the position of the patient, endoscopist and the endoscopic machine must be changed. However, in some situations, a supine position is advocated, such patients with extreme obesity, gross ascites and advanced pregnancy are often difficult to adopt the prone position.

Ferreira et al. [16] compared the outcomes between the supine position with the prone position, there were no differences between the 2 groups with respect to procedural time, the overall rates of success, complications or adverse cardiovascular events, but the degree of operation difficulty was significantly higher in the supine group. Mashiana et al. [17] reported their experience that the prone position has a higher surgical success rate with a slightly lower mean duration than the supine position, but the prone group has a higher number of adverse events of cardiopulmonary. The main difficulty is selective bile duct cannulation for therapeutic interventions in SIT patients. The European Society of Gastrointestinal Endoscopy (ESGE) published the clinical guidelines that percutaneous-guided approach can be used to achieve biliary pathway and antegrade guidewire insertion by ERCP when selective bile duct cannulation is unsuccessful [18]. To date, the percutaneous-transhepatic-endoscopic rendezvous procedure (PTE-RV) has been introduced more than 20 years, which combines percutaneous transhepatic cholangiography (PTC) with the endoscopic technique. Bokemeyer A [19] demonstrated that PTE-RV offer a high technical success rate (80%) in patients with a previously failed endoscopic retrograde cholangiography. At present, PTE-RV remains rescue approaches for biliary interventions.

In summary, keeping remember that the anatomy of SIT is mirrored from left to right, not from top to bottom. The endoscopist should be familiar with mirror symmetrical anatomy. The positions of ERCP are not standardized, but an appropriate position can be selected according to the patient's condition and practice of endoscopists. In addition, it is best to be operated by experienced endoscopists to ensure the success rate of operation and reduce the incidence of complications.

We report two cases in which a skilled endoscopist used two different positions. ERCP in SIT patients is generally safe and it will be easier in SIT patients with PTCD-guided rendezvous.

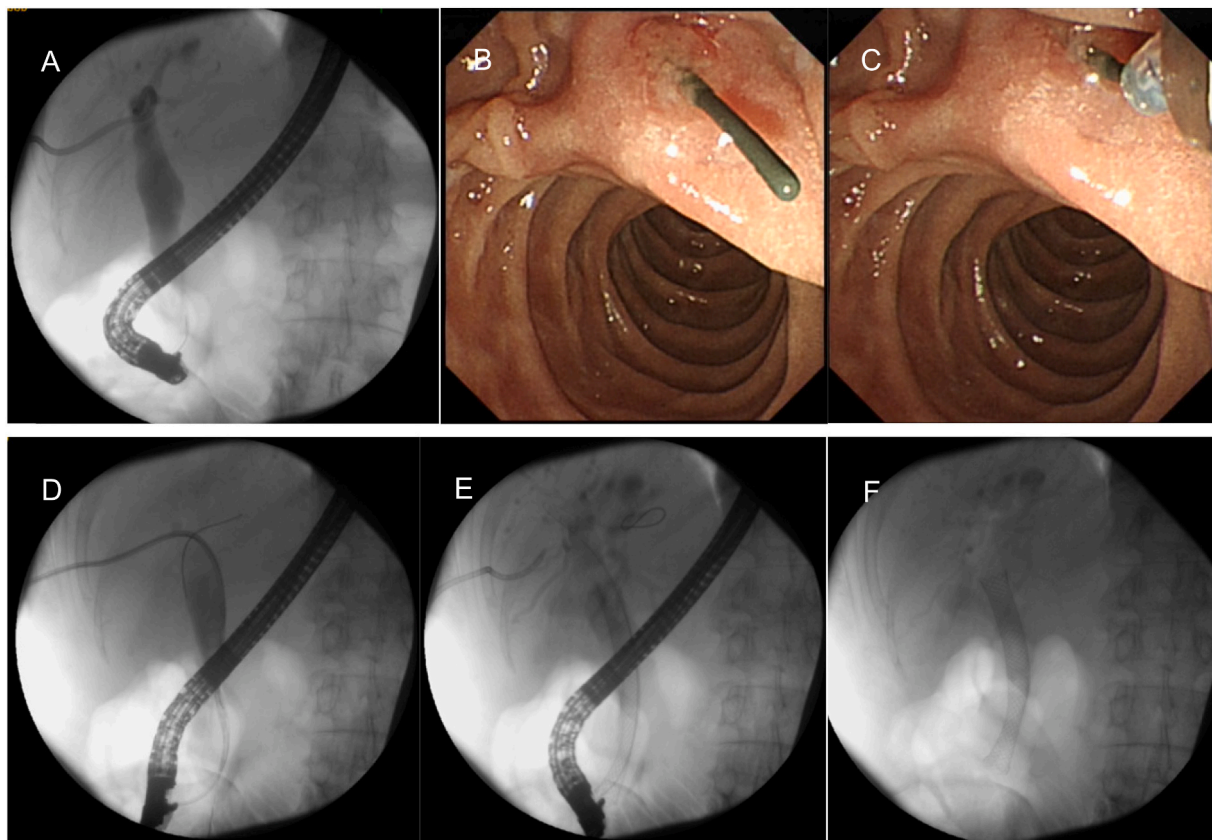


Fig. 7. A Showed the bile duct by injecting the contrast medium in PTCD. B Punctured the duodenal papilla by a guide wire through PTCD. C Docked the guide wire with a cutting knife under endoscope.

D Passed up the guide wire inside the cutting knife into the bile duct. E Released the EMBE under X-ray. F Reviewed the EMBE was fixed in place.

## Sources of funding

This research was supported by a grant of Science and Technology Project of Guangzhou City (grant number:202102010090).

## Ethical approval

The ethical committee approval was not required given the article type (case report).

## Consent

Written informed consent was obtained from the patients for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Registration of research studies

Not applicable.

## Guarantor

Liang Qi Cao.

## Provenance and peer review

Not commissioned, externally peer-reviewed.

## CRediT authorship contribution statement

All authors contributed in writing this paper.

## Declaration of competing interest

You Kui Gao, Song Hang Liu, Shui An Xie, Hao Peng Wen and Drs Liang Qi Cao have no conflicts of interest or financial ties to disclose.

## References

- [1] B. Casey, *Genetics of human situs abnormalities*, *Am. J. Med. Genet.* 101 (4) (2001) 356–358.
- [2] Y. Hu, H. Zeng, X.L. Pan, N.H. Lv, Z.J. Liu, Y. Hu, Therapeutic endoscopic retrograde cholangiopancreatography in a patient with situs inversus viscerum, *World J. Gastroenterol.* 21 (18) (2015 May 14) 5744–5748, <https://doi.org/10.3748/wjg.v21.i18.5744>. PMID: 25987802; PMCID: PMC4427701.
- [3] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, SCARE Group, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, *Int. J. Surg.* 84 (2020 Dec) 226–230, <https://doi.org/10.1016/j.ijsu.2020.10.034>. Epub 2020 Nov 9. PMID: 33181358.
- [4] T.H. Lee, S.H. Park, S.H. Lee, C.K. Lee, S.H. Lee, I.K. Chung, H.S. Kim, S.J. Kim, Modified rendezvous intrahepatic bile duct cannulation technique to pass a PTBD catheter in ERCP, *World J. Gastroenterol.* 16 (42) (2010 Nov 14) 5388–5390, <https://doi.org/10.3748/wjg.v16.i42.5388>. PMID: 21072905; PMCID: PMC2980691.
- [5] S. Sakazume, *Situs inversus viscerum, Ryoikibetsu Shokogun Shirizu* (34 Pt 2) (2001) 644–645. Japanese. PMID: 11528942.
- [6] T. Qiao, C. Yang, Endoscopic Retrograde Cholangiopancreatography for bile duct stones with residual cystic duct stones in patients with situs inversus totalis, *Asian J. Surg.* 43 (6) (2020 Jun) 709–710, <https://doi.org/10.1016/j.asjsur.2020.01.009>. Epub 2020 Feb 7 PMID: 32037056.
- [7] L. Hu, Y. Chai, X. Yang, Z. Wu, H. Sun, Z. Wang, Duodenoscope combined with laparoscopy in treatment of biliary stones for a patient with situs inversus totalis: a case report, *Medicine (Baltimore)* 98 (7) (2019 Feb), e14272, <https://doi.org/10.1097/MD.00000000000014272>. PMID: 30762727; PMCID: PMC6408073.
- [8] J.M. Lee, J.M. Lee, J.J. Hyun, H.S. Choi, E.S. Kim, B. Keum, Y.T. Jeon, H.J. Chun, H. S. Lee, C.D. Kim, Successful access to the ampulla for endoscopic retrograde cholangiopancreatography in patients with situs inversus totalis: a case report, *BMC Surg.* 17 (1) (2017 Nov 28) 112, <https://doi.org/10.1186/s12893-017-0307-x>. PMID: 29183311; PMCID: PMC5706404.
- [9] F. Feng, J. Yao, Hepatobiliary and pancreatic: common bile duct stones with situs inversus totalis, *J. Gastroenterol. Hepatol.* 35 (1) (2020 Jan) 12, <https://doi.org/10.1111/jgh.14833>. Epub 2019 Oct 24. PMID: 31412431.
- [10] A. Das, Performing an ERCP with the patient in the supine position: necessity is the mother of improvisation, *Gastrointest. Endosc.* 67 (7) (2008 Jun) 1044–1045, <https://doi.org/10.1016/j.gie.2007.12.012>. PMID: 18513547.
- [11] F.J. García-Fernández, J.M. Infantes, Y. Torres, F.J. Mendoza, F.J. Alcazar, ERCP in complete situs inversus viscerum using a "mirror image" technique, *Endoscopy* 42 (Suppl 2) (2010) E316–E317, <https://doi.org/10.1055/s-0030-1255813>. Epub 2010 Nov 26 PMID: 21113889.
- [12] I. Nordback, I. Airo, ERCP and endoscopic papillotomy in complete abdominal situs inversus, *Gastrointest. Endosc.* 34 (2) (1988 Mar-Apr) 150, [https://doi.org/10.1016/s0016-5107\(88\)71288-2](https://doi.org/10.1016/s0016-5107(88)71288-2). PMID: 3366336.
- [13] A. Chowdhury, B.K. Chatterjee, U. Das, P. Dutta, G.K. Dhali, P.K. Banerjee, ERCP in situs inversus: do we need to turn the other way? *Indian J. Gastroenterol.* 16 (4) (1997 Oct) 155–156. PMID: 9357192.
- [14] F. Fiocca, G. Donatelli, V. Ceci, F. Cereatti, F. Romagnoli, L. Simonelli, C. Modini, ERCP in total situs viscerum inversus, *Case Rep. Gastroenterol.* 2 (1) (2008 Mar 14) 116–120, <https://doi.org/10.1159/000119713>. PMID: 21490849; PMCID: PMC3075177.
- [15] C. de la Serna-Higuera, M. Perez-Miranda, G. Flores-Cruz, P. Gil-Simón, A. Caro-Patón, Endoscopic retrograde cholangiopancreatography in situs inversus partialis, *Endoscopy* 42 (Suppl 2) (2010) E98, <https://doi.org/10.1055/s-0029-1214968>. Epub 2010 Mar 19. PMID: 20306407.
- [16] L.E. Ferreira, T.H. Baron, Comparison of safety and efficacy of ERCP performed with the patient in supine and prone positions, *Gastrointest. Endosc.* 67 (7) (2008 Jun) 1037–1043, <https://doi.org/10.1016/j.gie.2007.10.029>. Epub 2008 Jan 18 PMID: 18206877.
- [17] H.S. Mashiana, M. Jayaraj, B.P. Mohan, G. Ohning, D.G. Adler, Comparison of outcomes for supine vs. prone position ERCP: a systematic review and meta-analysis, *Endosc. Int. Open* 6 (11) (2018 Nov) E1296–E1301, <https://doi.org/10.1055/a-0603-3302>. Epub 2018 Nov 7. PMID: 30410948; PMCID: PMC6221825.
- [18] P.A. Testoni, A. Mariani, L. Aabakken, M. Arvanitakis, E. Bories, G. Costamagna, J. Devière, M. Dinis-Ribeiro, J.M. Dumonceau, M. Giovannini, T. Gyokeres, M. Hafner, J. Halttunen, C. Hassan, L. Lopes, I.S. Papanikolaou, T.C. Tham, A. Tringali, J. van Hooft, E.J. Williams, Papillary cannulation and sphincterotomy techniques at ERCP: European Society of Gastrointestinal Endoscopy (ESGE) clinical guideline, *Endoscopy* 48 (7) (2016 Jul) 657–683, <https://doi.org/10.1055/s-0042-108641>. Epub 2016 Jun 14 PMID: 27299638.
- [19] A. Bokemeyer, F. Müller, H. Niesert, M. Brückner, D. Bettenworth, T. Nowacki, T. Beyna, H. Ullerich, F. Lenze, Percutaneous-transhepatic-endoscopic rendezvous procedures are effective and safe in patients with refractory bile duct obstruction, *United Eur. Gastroenterol. J.* 7 (3) (2019 Apr). Epub 2019 Jan 17. PMID: 31019708; PMCID: PMC6466745.