



Research Article

ENDOSCOPIC RETROGRADE CHOLANGIOPANCREATICO GRAPHY IN SITUS INVERSUS TOTALIS

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ABSTRACT

Situs inversus is a rare congenital anomaly with transposition of viscera in which the internal organs are reversed, as if in a mirror image of their normal position.. Endoscopic retrograde cholangiopancreatography (ERCP) in patients with situs inversus because of mirror image location of Upper GI tract and biliary system. Many positions and approaches are described for performing ERCP. However, we have performed ERCP in in conventional position without much difficulty, and we report two cases of situs inversus totalis who underwent ERCP in standard prone position and this is the First case report in patient who underwent Gastrojejunostomy (GJ)

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INTRODUCTION

Situs inversus is a rare congenital anomaly with a genetic predisposition with an Autosomal recessive inheritance¹. Situs inversus is due to mutation in long arm of chromosome 14 resulting in rotation of midgut loop in clock wise rather than counter clock wise direction² during embryonic period, with a frequency of 1:5,000 to 1:10,000 live births. Fabricius described for the first time situs viscerum inversus in humans in this condition the organs are transposed either, totally or partially, to the opposite side of the body.

Endoscopic sphincterotomy and stone extraction are standard procedures for the removal of common bile duct stones. It is more difficult to perform endoscopic retrograde cholangiopancreatography (ERCP) and sphincterotomy in patients with situs inversus than in normal patients^{3,4} because of altered Reverse anatomy

Endoscopic retrograde cholangiopancreatico graphy (ERCP)

ERCP is a procedure done with a side viewing endoscope and the patient was kept on prone position. In Situs Inversus Totalis there is a complete transposition of all viscera, dextroposition of the heart, and the liver is situated on the left side. The anatomy of the left and right sides is reversed. ERCP is technically challenging in these pts because of mirror image location of Upper GI tract and biliary system, normal conventional position is not recommended as the papilla can be cannulated in long route only¹ that ERCP should be performed in supine position.

Although various endoscopic techniques in situs inversus have been reported. The side viewing endoscope, is built for the right position of the liver with the papilla at the medial side of the duodenum. Accessories used in ERCP are built to use in the normal direction of the biliary tract. ERCP in patients with situs inversus is always challenging even for an experienced Endoscopist.

Till now, There are 10 published cases till 2016. However there are, no case reports of ERCP in situs inversus pt with GJ. We report two cases of situs inversus totalis who underwent ERCP in standard prone position case reports of ERCP in pt with Gastro jejunostomy (GJ). We performed ERCP in conventional position and we believe ERCP can be performed in conventional position with small changes in the maneuvers and Endoscopic tip movements.

Case 1: A 45 yr old female who underwent GJ 18 yrs back presented with recurrent epigastric pain of 18 months duration and Jaundice, fever and pruritus of 1week .On physical examination she is febrile, and tenderness of epigastric region. ERCP and Endoscopic sphincterotomy performed in conventional position successfully.

Investigations: WBC - 6,000/cc, Hb% -10gms%, ESR -14 & 24 mm/hr. S.Amylase -30 S.U/dl, S.Bil - 2.8 mg/dl (0.3-0.8), SGPT-200 IU/dl (20-93) Alk.phos – 60 KaU/dl (3-13), PT-16/13sec. Chest X ray revealed Dextrocardia 2DECHO - revealed normal cardiac function

U/Sound scan of abdomen confirmed situs inversus totalis, with 15mm calculus in distal CBD and IHBD. Upper GI endoscopy-GJ stoma seen and duodenum is Normal.

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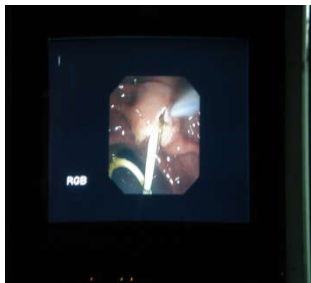
ERCP was performed with the pt in the conventional left lateral decubitus position and the Endoscopist on the left side of the table. The antrum; pylorus and first part of duodenum were crossed without difficulty. By turning to left scope is advanced into descending duodenum. Papilla made enface by counter clock wise rotation (left 90 °) and withdrawing the scope. Initially MPD cannulated at 12 O'clock position. CBD is cannulated With difficulty at 2o'clock position CBD is dilated with a single stone at hilum 8cm, 8F double pig tail stent inserted and ES done over the stent with NKS. After stenting pain subsided LFT became normal. She underwent cholecystectomy and CBD exploration



ERCP in situs with GJ



Cholangiogram with CBD



CBD stent in situs Pt

Case 2: A40 yrs old middle aged male known case of with situs inversus presented with pain abdomen and jaundice of 7 days duration.

Investigations: WBC - 7600/cc, Hb% -13gms%, Amylase -80 S.U/dl, S.Bil - 1.8 mg/dl (0.3-0.8), SGPT – 180 IU/dl (20-93) Alk.phos – 82 KaU/dl (3-13), PT- 16/13sec

U/S reveal GB stones with normal CBD. In view of Elevated Liver enzymes, ERCP attempted in conventional position and ES done 7fr stent placed in CBD and after laparoscopic cholecystectomy was done.

DISCUSSION

Although widely used in patients with biliary tract disease, ERCP procedures are challenging in the presence of anatomical abnormalities and even a skilled Endoscopist can encounter technical difficulties when performing ERCP in

patients with situs inversus. There have been only a few reports on ERCP in patients with situs inversus [2, 3, 4, 5, 6].

Difficulties are

1. Access to duodenal papilla.
2. Maneuverability of Scope
3. Use of accessories designed for normal anatomy
4. Post-operative pts with altered anatomy

Biliary cannulation has a success rate of almost 100% with the patients in the prone position⁴. Many reports showed that the supine position does not affect the feasibility of ERCP when the operator turns the instrument clockwise 180 degrees⁵

1. Position of the patient
2. Position of the Endoscopist
3. Maneuvers during Procedure

Position of the Patient

Prone position: Although the patient lies prone and the Endoscopist performs ERCP on the right side of the table, as in normal patients, several techniques have been used in situs inversus to increase the success rate [4, 5, 6].

Supine position: In Supine position, ERCP will be difficult for three reasons: first, the head could not be fixed easily in a natural position; second, cannulation was difficult when the papilla sank into the intestinal wall because of the effect of gravity and the diverticulum, which increased the difficulty of intubation and likelihood of papillary edema and damage to the pancreatic duct. In addition, the stomach could not be fixed for stability of the endoscope in the supine position. In general, a supine position was adopted for duodenoscopic examination because the anatomical view remains consistent; however, the traditional prone position, in which the endoscope needs to be rotated clockwise 180°, can be applied for maintenance and cannulation in patients with SIV,

Position of the Endoscopist: Endoscopist performs the procedure from the left side or right side of the patient

Maneuvers during Procedure

In subjects with Situs inversus, a mirror image of the normal anatomy makes it harder for Endoscopist's to gain access to duodenal papilla. From the cases reported, most of the patients were placed in a left lateral position or prone position, which is the usual position in ERCP, and the Endoscopist preferred to be at the right side of the operating table. Adjustments such as a 180° turn in the stomach or special techniques such as a "mirror imaging" were shown to contribute to maintenance of the duodenal papilla.

The Twist method: The patient was placed in the prone position and the Endoscopist's performed ERCP from the right side of the table. In The "twist" method, the endoscope was rotated 180° clockwise in the stomach¹⁶. After entering the duodenum, the endoscope was again shortened using a 180° counterclockwise rotation although access was relatively easy, the ampulla in the endoscopic view was deviated to right side and right-upward direction Moreover, it was difficult to control the endoscope owing to the looped endoscope shaft¹⁷.

The "mirror image" method. The patient is usually placed in the right prone position because of the reversal of internal organs, and the Endoscopist performs the procedure from the left side of the table. However, shortening the endoscope using a counterclockwise rotation in the mirror image method is

inconvenient, as the Endoscopist is required to manipulate the endoscope with the right hand. In addition, the position of the patient, Endoscopist, the endoscopic machine, and the monitor must be changed.

The “inversed normal “method: ERCP can be performed with the patient in the prone position and the Endoscopist on the right side of the table. The endoscope is advanced to the duodenum by following the lesser curvature without rotating the endoscope.

To date, nine cases of therapeutic ERCP in patients with Situs inversus totalis have been reported worldwide (Table - 1).

A synopsis of Published case reports on ERCP positions in patients with situs inversus totalis

Author-ref	year	Patient position	Endoscopist position	Maneuvers	Complications	Difficulties
Venu etal (USA) ⁷	1985	Right lateral	Right side	Pt position altered during procedure	Nil	
Nordback ⁸ (Finland)	1988	Right Prone	----	Endoscopist position altered	Nil	
Chowdery et al ⁹ (India)	1997	Left lateral	Left side	-	Nil	
Fiocca et al ¹⁰ (Italy)	2008	prone	Right side	180 Degrees turn	Nil	Difficulty in reaching duodenum
Garcia-Fernandez (Spain) ¹¹	2010	Right lateral	Right side	Inverse movements	Nil	
De la serna – Higuera(Spain) ¹²	2010	Prone	Right side	Rotatory movements	Nil	Difficulty in reaching duodenum
Lee et al ¹³ (south Korea)	2010	Prone	Right side	180°rotation	Nil	Difficulty in reaching duodenum
Kamani et al ¹⁴ Pakistan	2014	Left lateral	Right side	180°rotation	Nil	

ERCP in Other situations in patients with situs Inversus

Post-Operative status: ERCP can be technically challenging because of anatomic variants, when operative alteration are carried out on the upper digestive system (such as Bill Roth I, Bill Roth II gastric resection, Roux-en-Y enteral anastomosis etc.)². Endoscopic access to the papilla is 92% in Bill Roth II and 33% in Roux-en-Y reconstructions³. **However there are No case reports of ERCP in pt with Gastro jejunostomy (GJ)**

In Chronic Pancreatitis: There are case reports of ERCP being done in pts with chronic pancreatitis in whom passage of scope into duodenum is facilitated by turning the pt to right side and papilla is cannulated in long route¹⁵. In one pt of situs with pseudo pancreatic cyst, ERCP performed in conventional position and Endoscopist turning to left side¹⁸ however performing a technically difficult procedure in an unaccustomed position is difficult and may reduce the success rate

Periampullaery diverticulum

CBD cannulation is more difficult in patients with a Periampullaery diverticulum.

In this we report an unusual case of CBD stones in a patient with situs inversus with GJ. However there are No case reports of ERCP in pt with Gastro jejunostomy (GJ). Hence we, started ERCP in conventional position and we believe ERCP can be performed in conventional position with small changes in the maneuvers and Endoscopic tip movements. Successful ERCP was performed using conventional technique, as the ampulla was located at the center of the screen, slightly to the left. However, insertion of the endoscope was relatively

difficult. Real understanding of anatomy is the key success factor in this unique case. In conclusion, this report shows that it is possible to perform ERCP in patients with anatomical alterations condition, and also in patients with situs inversus totalis.

CONCLUSION

In conclusion, therapeutic ERCP is a safe procedure for patients with Situs inversus Totalis. However, ERCP can be performed more smoothly with conversion of the patient’s position or the use of special techniques. The major emphasis in ERCP is adjusting the patient and Endoscopist positions during the operation. We believe ERCP can be performed in conventional position with small changes in the maneuvers and Endoscopic tip movements.

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