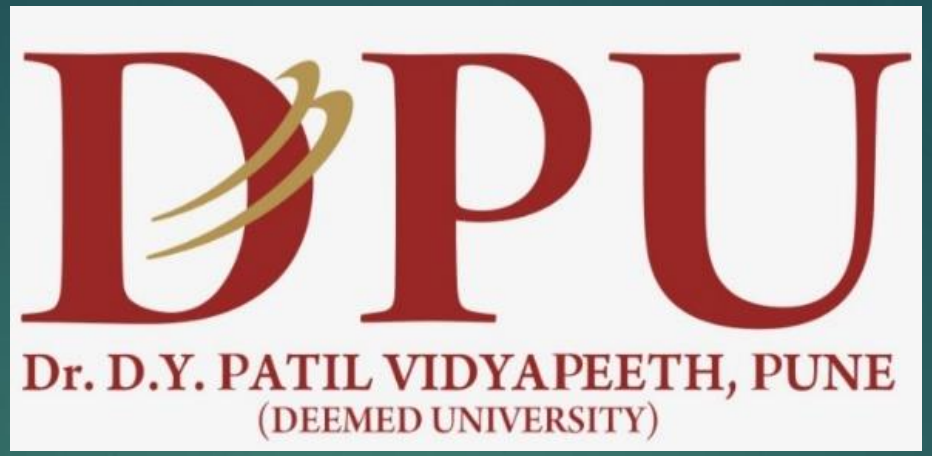

LEMMEL SYNDROME – AN ATYPICAL PRESENTATION OF A RELATIVELY TYPICAL DIAGNOSIS

UNIT IV

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INTRODUCTION

- Intestinal diverticulosis is an uncommon occurrence with most cases being diagnosed incidentally.
- **Duodenal diverticuli are relatively more frequent than jejuno-ileal diverticuli**.
- Amongst these , periampullary diverticula are those that occur within 2-3 cms of the ampulla
- Usually quiescent, periampullary diverticula can occasionally become inflamed and have a wide plethora of presentation
- **Lemmel Syndrome signifies distal CBD obstruction due to external pressure by a peri-ampullary diverticulum in the absence of choledocholithiasis**
- We describe a rare case of obstructive jaundice brought on by extrinsic compression by a large periampullary diverticulum that was successfully treated with surgical intervention.

CASE REPORT

- A 60 years old female presented with chief complaints of :
  - **Pain in the upper abdomen since last 6 months**
  - **Yellow discoloration of eyes since 3 months.**

CASE REPORT

- **History of Presenting Illness**:

- Patient was apparently normal 6 months back.
- Pain in the **right upper abdomen** for 6 months.
  - Insidious in onset - Gradually progressive in nature
  - **Dull aching** in nature - Non-radiating
  - Aggravated with meals and relieved with medications.
- H/O **Jaundice** since 3 months
  - Insidious in onset with **gradual progression**.
  - No H/O waxing and waning of jaundice.
  - Not associated with pruritis..

CASE REPORT

- H/O multiple episodes of vomiting – **Nonbilious, post meals**.
- H/O loss of appetite
- No H/O fever/ weight loss.
- No H/O similar complaints in the past.

❖ **Past History:-**

- No H/O DM/HTN/Asthma.
- No H/O previous surgeries.

CASE REPORT

- **Personal History:**
  - Mixed diet
  - Normal bowel and bladder movements
  - No addictions

- **Family History**: Not significant.

CASE REPORT - GENERAL EXAMINATION

- Patient was conscious, coherent, and cooperative – Oriented to time place, and person
- BMI: 24 kg/m^2
- Icterus +
- Moderately built and nourished.
- Afebrile
- PR: 86/min
- BP:110/80 mm of Hg

CASE REPORT – PER ABDOMEN EXAMINATION

- **Inspection:**
- Abdomen is obese.
- Umbilicus - Centrally placed and inverted.
- All quadrants of the abdomen move equally with respiration.
- No visible fullness or lump – **No visible peristalsis.**

- **Palpation**:
- **Tenderness was present in the epigastric region and right hypochondrium**.
- **No palpable abdominal lump**.
- No organomegaly.

CASE REPORT – PER ABDOMEN EXAMINATION

- **Percussion**: No free fluid in the abdomen.
- **Auscultation**: Bowel sounds present in all quadrants – Normal in nature

COURSE IN HOSPITAL

❖ On admission, routine haematological and biochemical investigations were performed.

BLOOD PARAMETER		VALUES (ON ADMISSION)
Haemoglobin		9.8 g/dl
Total leucocyte count (TLC)		**12000/mm^3**
Liver function test (LFT)	Serum bilirubin (Total)	**8 mg/dl**
	Serum bilirubin (Conjugated)	**6 mg/dl**
Renal function test (RFT)		Within normal limits
Alkaline Phosphatase levels (ALP)		
Serum amylase/ Serum Lipase		**Within Normal Limits**
PT/INR		16/1.2

- Ultrasound (Abdomen/Pelvis):

- **Dilated CBD (9mm at the porta hepatis) with oedematous head of pancreas and small GB calculi**
- No free fluid in abdomen.

- Contrast-enhanced computed tomography (CECT - Abdomen and Pelvis):

- **A periampullary diverticulum (4*3*4 cms) arising along the posteromedial aspect of 2nd part of the duodenum + GB calculi**
- **The diverticulum appears to be compressing the distal CBD with proximal CBD/CHD dilatation.**
- Asymmetric thickening of the pyloric region of the stomach (?Gastritis)

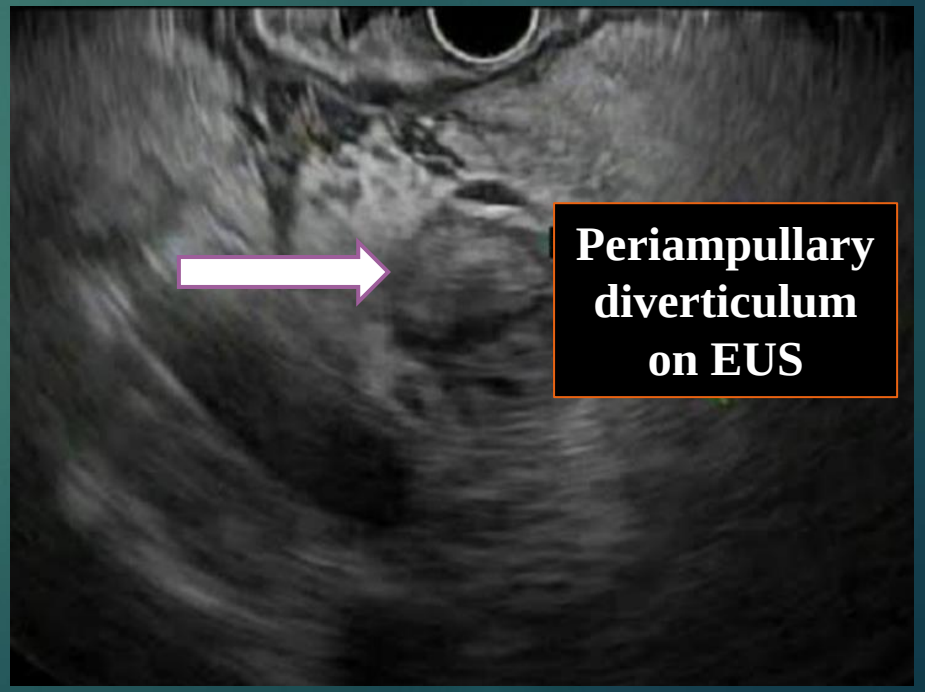
COURSE IN HOSPITAL

- **Magnetic resonance cholangiopancreatography (MRCP) :**

- Narrowing at the distal CBD with proximal dilatation of CBD/CHD with Gallbladder calculi.
- Features s/o Lemmel syndrome on correlation with CECT (A/P)

MRCP showed dilated CBD in the absence of choledocholithiasis/ Neoplasm

- **Endoscopic Ultrasound:**
  - **Periampullary diverticulum 4*3*3cms**
  - **Dilated CBD** – Maximum diameter 10.2mm
  - Gallbladder calculi
  - **No e/o choledocholithiasis**



Periampullary diverticulum on EUS

COURSE IN HOSPITAL

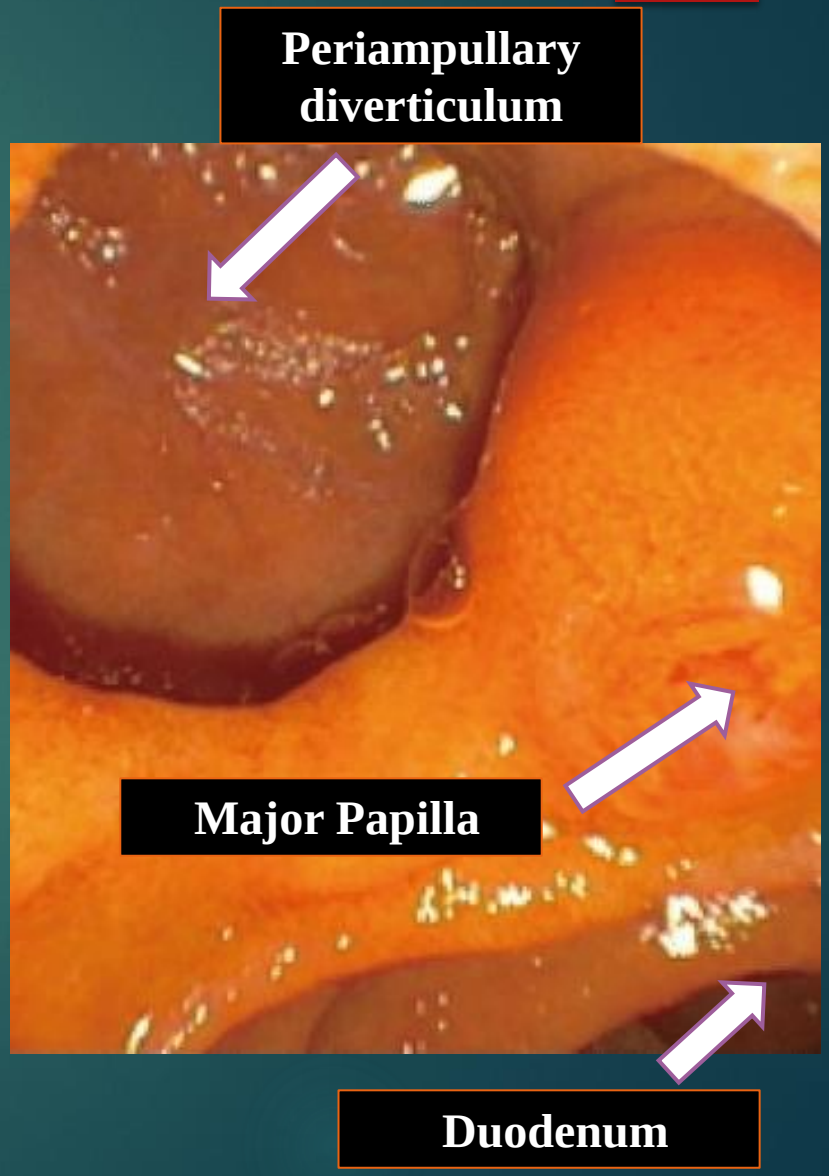
- **Upper GI endoscopy** was performed considering the symptomatology and radiological findings: **Chronic Antral gastritis with no mass lesions**

  - Rapid Urease Test : Negative

- An **ERCP** was then planned to facilitate stenting of the distal CBD and the pancreatic duct. However, despite 2 attempts the procedure could not be performed **due to friable mucosa and bleeding.**

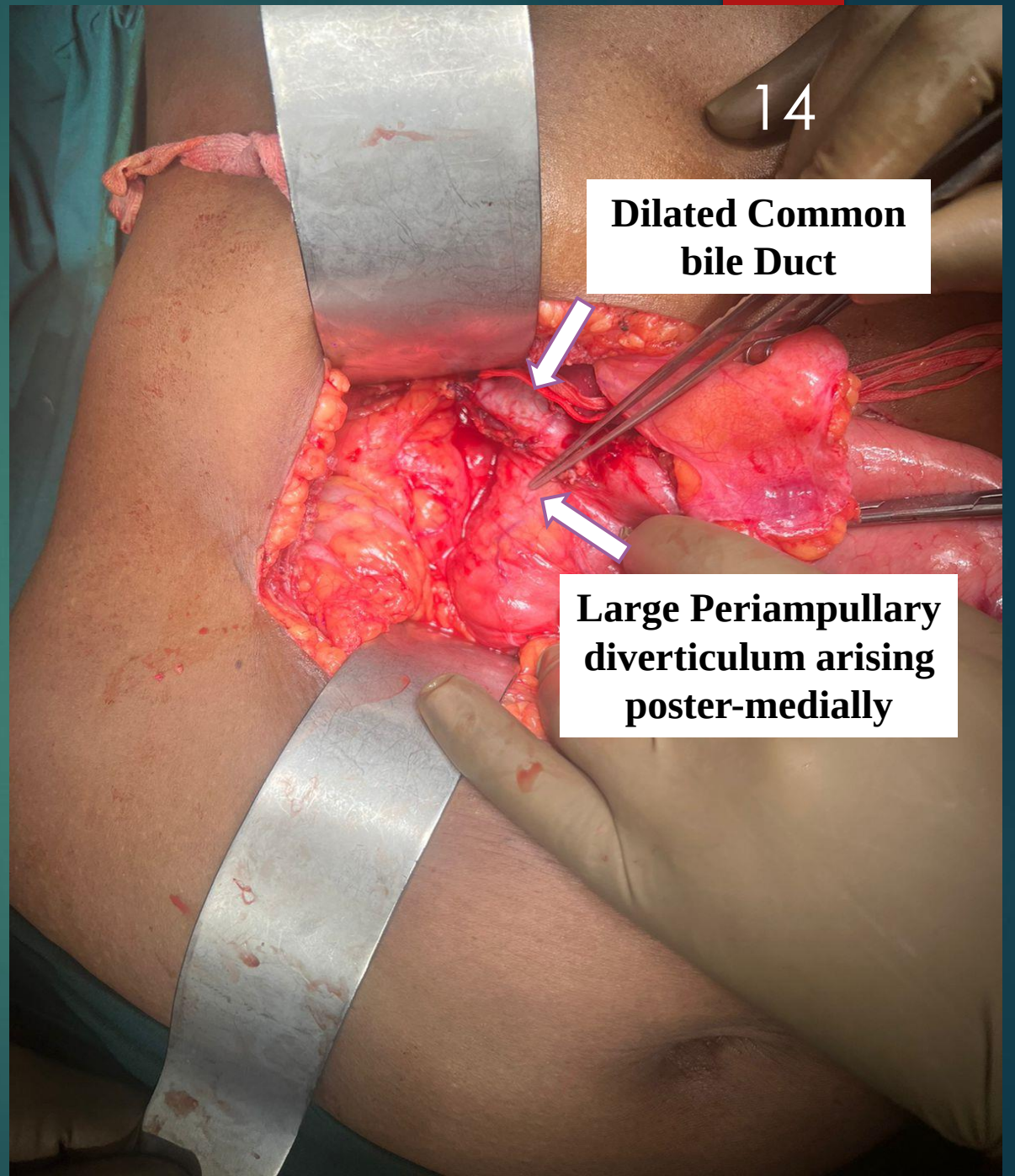
- With all investigations exhausted we decided to go ahead with surgical intervention .

- After adequate pre-operative optimization, and anasthetic fitness the patient was taken for an Exploratory Laparotomy.



COURSE IN HOSPITAL

- **Incision** : Chevron Incision (Rooftop incision)
- On exploration and adequate Kocherisation of the duodenum, a **4*3 cm diverticulum was identified and arose from the postero-medial wall of the 2nd part of the duodenum.**
- **Cholecystectomy was performed**

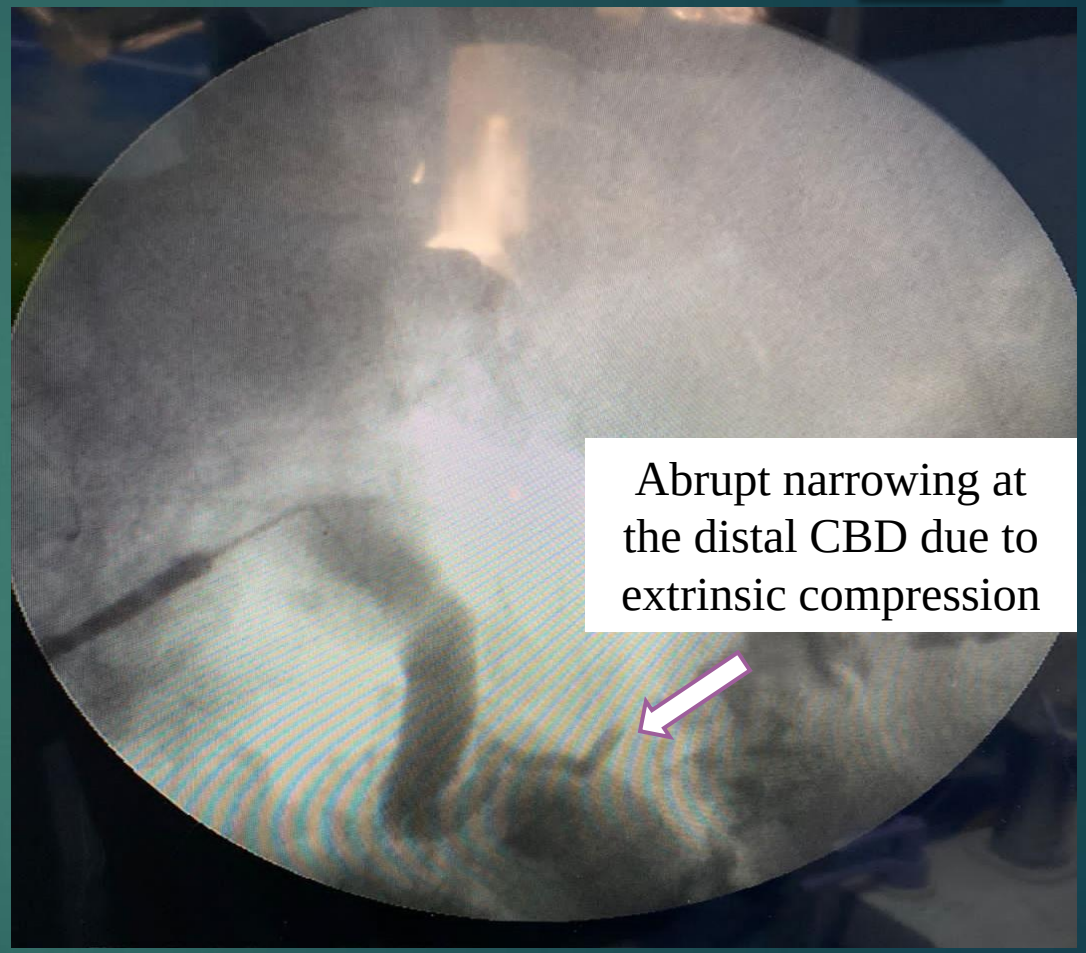


COURSE IN HOSPITAL

- Intraoperative cholangiogram:

Abrupt narrowing at the distal CBD with dye reaching into the duodenum indicating partial obstruction.

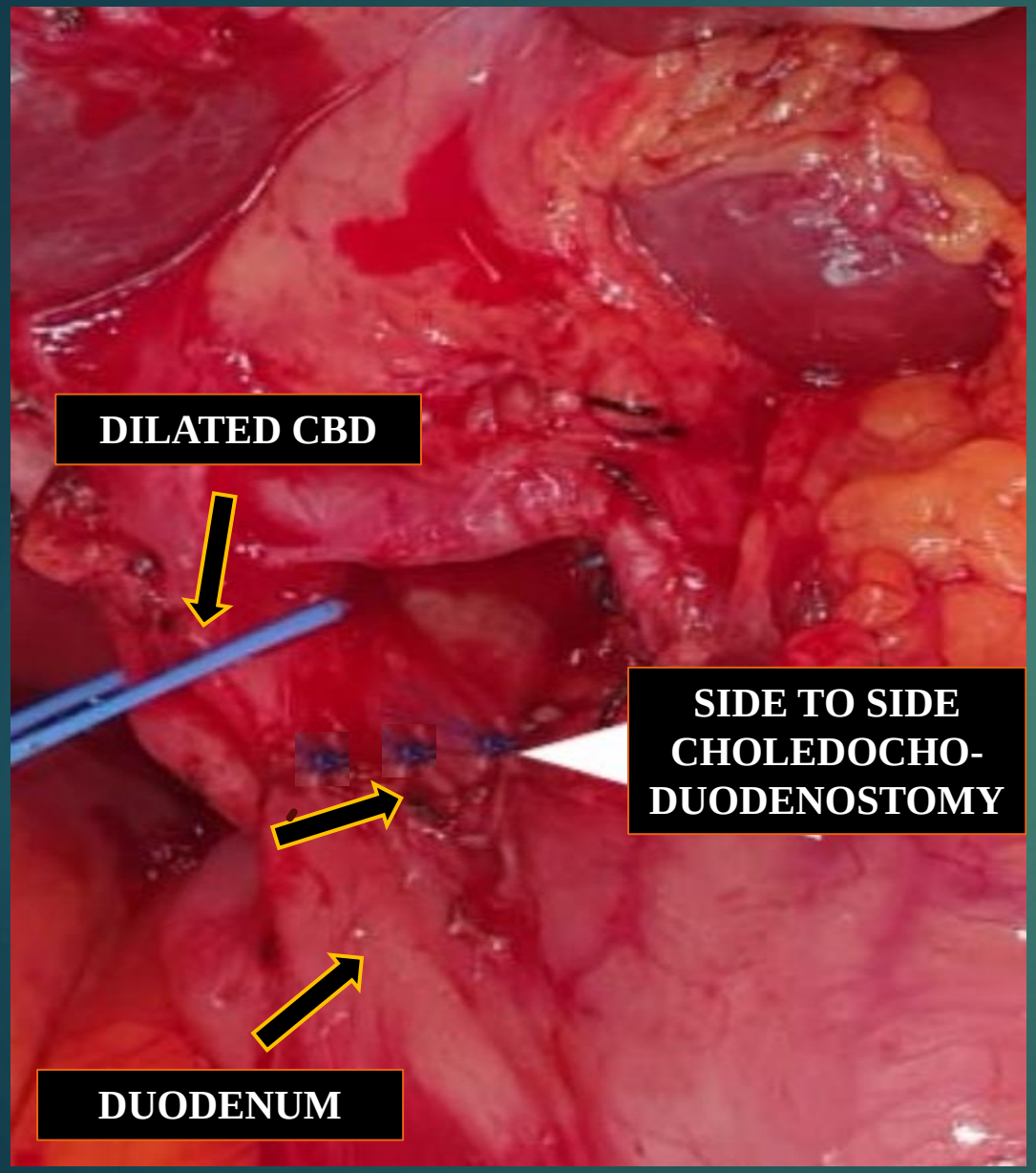
- Due to posterior origin of the diverticulum and extensive inflammation associated with the diverticulum , a bypass procedure was planned.

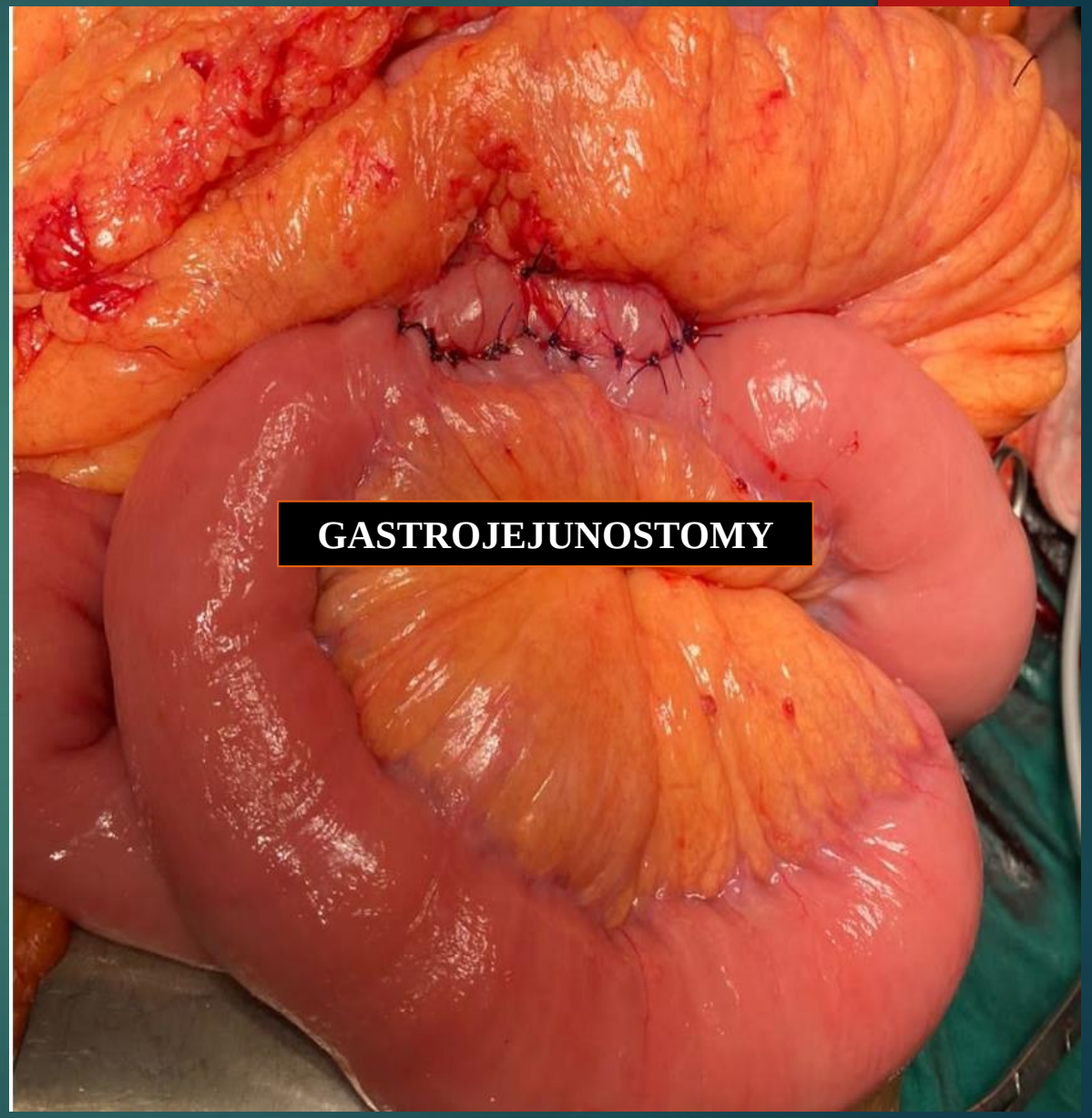


COURSE IN HOSPITAL

- **Isoperistaltic Retro colic Posterior Gastrojejunostomy and Side to side Choledocho-Duodenostomy was performed.**
- The procedure was uneventful and the patient had a smooth postoperative recovery.
- **On the review visit after 3 months, the patient was symptom-free and follow-up blood work showed a drastic improvement in the previously deranged parameters**

PRE –OPERATIVE BILIRUBIN LEVELS	POST-OPERATIVE BILIRUBIN LEVELS
Conjugated Hyperbilirubinemia	Normal LFT's
Sr.Bilirubin (Conjugated): 6mg/dl	Sr.Bilirubin (Conjugated): 0.8mg/dl


DILATED CBD
SIDE TO SIDE CHOLEDOCHO-DUODENOSTOMY
DUODENUM


GASTROJEJUNOSTOMY

DISCUSSION

- **Periampullary diverticula constitute the most common type of duodenal diverticula accounting of ~ 12 %.**
- **In most instances, periampullary diverticuli remain asymptomatic**, but up to 1% of them may be exacerbated by diverticulitis, haemorrhage, perforation, obstructive jaundice (Lemmel syndrome), choledocholithiasis, pancreatitis, or cholangitis.
- **Obstructive jaundice though rare usually occurs a result of extrinsic compression of the common bile duct (CBD) by the diverticulum. This phenomenon is known as Lemmel Syndrome and carries an incidence of ~2%**

DISCUSSION

- **Pathophysiology of Lemmel syndrome:**

- Firstly, primary irritation of the ampulla by the inflamed diverticulum can cause fibrosis and scarring of the ampulla with resultant jaundice.
- Secondly, the sphincter of Oddi may become dysfunctional as a result of the periampullary diverticula.
- Thirdly, periampullary diverticula, as in our instance, can mechanically compress the distal CBD or ampulla.

DISCUSSION

- **Lemmel Syndrome remains a diagnosis of exclusion** in a case of surgical obstructive jaundice.
- An extensive battery of investigations is required to establish the diagnosis, of which **EUS followed by MRCP are the most sensitive** (Diagnostic accuracy ~ 80% and 75% respectively)

DISCUSSION

- The management of Lemmel Syndrome varies from and is not limited to :
  - Conservative management in mildly symptomatic cases with obstructive jaundice.
  - Endoscopic management i.e ERCP +/- Sphincterotomy SOS CBD stenting.
  - Open/Laparoscopic surgery in unresponsive and resistant cases i.e
    - ~ Diverticulectomy (Technically challenging with high mortality ~ 2%)
    - ~ **Bypass procedure (Gastrojejunostomy with Choledocho-Enteric anastomosis)**

UNIQUE POINTERS IN THIS CASE

- **Amongst the wide plethora of causes for obstructive jaundice, Lemmel syndrome constitutes only 0.2% and remains a diagnosis of exclusion in order to avoid mis/undertreatment.**
- **The diagnosis of Lemmel syndrome requires a high index of suspicion along with detailed understanding of periampullary anatomy.**
- **Management requires a multi-disciplinary approach with surgical options reserved in resistant cases.**

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THANK YOU