Interhospital Conference

โรงพยาบาลจุฬาลงกรณ์

พญ. พลิตถิยา สินธุเสก

นพ. อนันดร วงศ์ธีระสุต

พญ. กิตติยา เศรษฐไกรสิงห์

ผู้ป่วยเด็กชายไทย อายุ 4 ปี

- ภูมิลำเนา กทม.

- CC : ปวดท้องเป็นๆหายๆ นาน 3 เดือน

ผู้ป่วยเด็กชายไทย อายุ 4 ปี

ประวัติปัจจุบัน

- 3 เดือนก่อนมาร.พ. : ปวดท้องบริเวณรอบๆสะดือ ไม่มีปวดร้าว ปวดแน่นๆนานประมาณ 15-20 นาทีต่อครั้ง วันละ2-3ครั้ง มีอาการปวด 3-4 วัน ปวดเป็นๆหายๆ ไม่สัมพันธ์กับอาหาร ห่างกันประมาณ 2-3 อาทิตย์ ไม่มีปวดตอนกลางคืน มีคลื่นไส้อาเจียนไม่มีไข้ ไม่มีอาการตัวเหลือง ปัสสาวะสีไม่เข้ม อุจจาระไม่ซีด ไปตรวจที่โรงพยาบาลเอกชน 3 ครั้ง ได้ยาลดอาการอาเจียน ยาแก้อักเสบรับประทาน อาการดีขึ้น

ผู้ป่วยเด็กชายไทย อายุ 4 ปี

- **1 สัปดาห์** : ปวดท้องมากขึ้น ปวดนานเป็นชั่วโมง ญาติสังเกตุว่าตาเหลืองมากขึ้น ปัสสาวะสีเข้มกว่าเดิม ถ่ายอุจจาระสีซีดลง ไม่มีอาการคลื่นไส้อาเจียน ไม่มีไข้ ไปตรวจโรงพยาบาลเอกชน แพทย์บอกเป็นตับอักเสบจึงส่งตัวมารักษาต่อที่ร.พ.จุฬา

  CBC : Hct 38.6, Hb 13, WBC 7,600 (P46, L46),
  Plt 277,000

  LFT : Alb 4.3, Glob 3.4, TB 4.1, DB 3.2,
  AST 607, ALT 579, AP 556, GGT 661

ประวัติอดีต

- ไม่มีประวัติตัวเหลืองตาเหลืองมาก่อน
- ไม่มีประวัติแพ้ยา ไม่ได้ใช้ยาชนิดใดๆเป็นประจำ
- ไม่มีประวัติได้รับเลือด
- ประวัติวัคซีนครบตามกำหนด
- ไม่มีโรคประจำตัว

ประวัติการคลอด

- บุตรคนที่ 1/1
- คลอด C/S due to fetal distress
- น้ำหนักแรกคลอด 3,750 กรัม
- หลังคลอดแข็งแรงดี ไม่มีประวัติตัวเหลือง

ประวัติครอบครัว

- ไม่มีประวัติโรคประจำตัวในครอบครัว
- ไม่มีประวัติโรคเลือด, โรคโลหิตจางในครอบครัว

Physical examination

Physical examination

- **Vital sign** : T 36.4 °C, HR 70/min, RR 20/min, BP 90/65

  BW 17 kg (P50-75) , Height 106 cm (P75-90)

- **GA** : Thai boy, active, afebrile, mild jaundice, not pale
- **HEENT** : mild icteric sclera, not pale conjunctiva, no cervical lymphadenopathy, tonsils not enlarged
- **Lung** : normal breath sound, no adventitious sound
- **CVS** : full & regular pulse, no murmur

Physical examination

- **Abdomen** : mild distension, no superficial vein dilatation, mild tender at periumbilical region, no guarding & rebound, no palpable mass, liver 1 cm below RCM (Span 6 cm), soft and smooth surface, spleen not palpable
- **Extremities** : no rash, no ecchymosis

PROBLEM LISTS

Problem lists

- **Chronic abdominal pain**
- **Jaundice**
- **Hepatitis**

Differential diagnosis

DDX

- Choledochal cyst
- Gall stone
- Acute viral hepatitis
- Tumor

- Infection : parasite, …
- Tumor
- Anomaly : choledochal cyst

Investigation

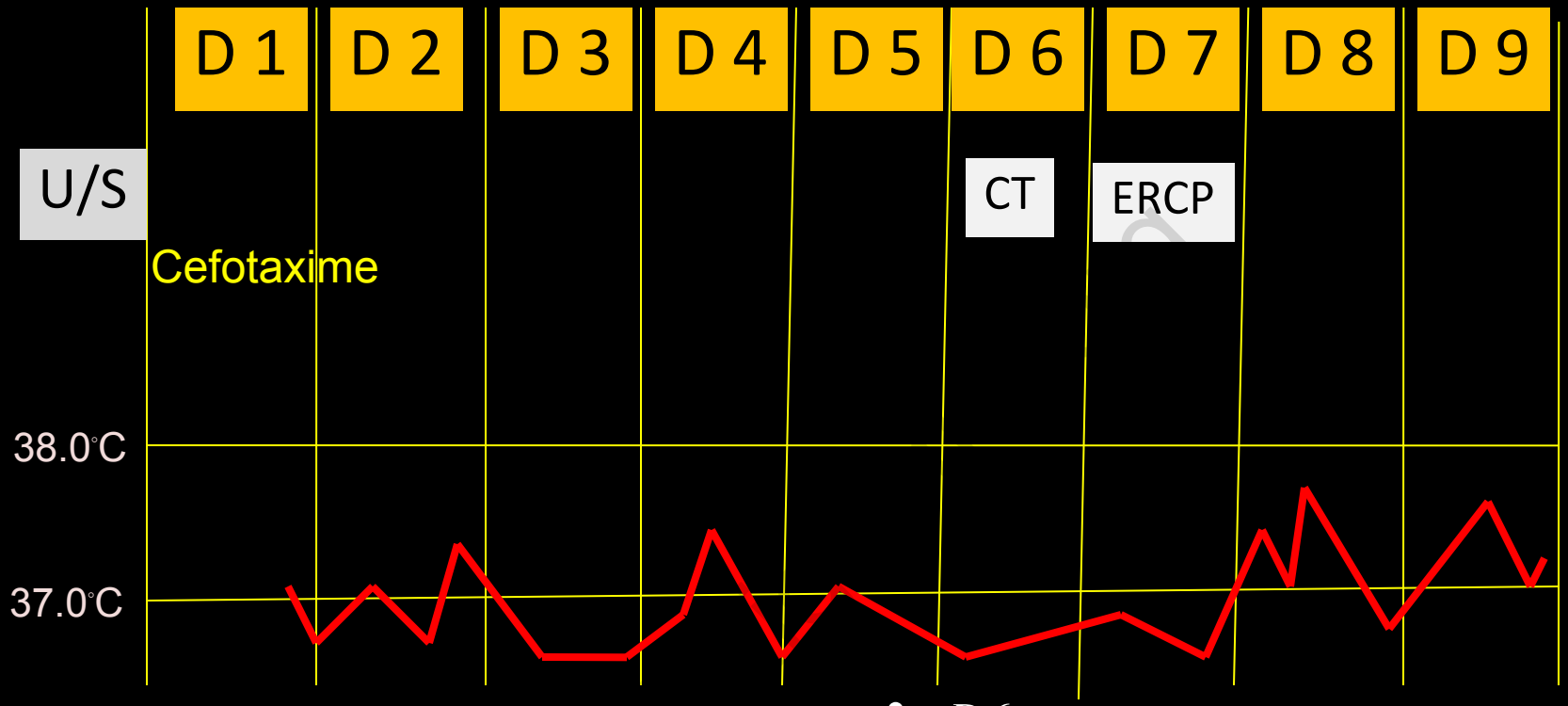
1 2 3 4

5 6 7 8

9 10 11 12

Treatment

- NPO
- IV fluid
- Pethidine 15mg IV prn q 8 hr
- Cefotaxime (100mg/kg/day)



- D 1
- LFT : Alb 4.6 , Glob 3.4 ,
  AST 345 , ALT 647,
  TB 4.03 , DB 3.35 , AP 678
- Amylase 565, Lipase 1696

- D 6
- LFT : Alb 4.1 , Glob 2.7,
  AST 49 , ALT 170 ,
  TB 0.93 , DB 0.78 , AP 386
- Amylase 139, Lipase 243

D 8

- Total cholesterol 251, HDL 36, Triglyceride 101, LDL 176
- Calcium 9.8, Phosphate 4.4
- LFT : Alb 4.2, Glob 3.1 g/dl, TB 0.91 , DB 0.81 mg/dl,
  AST 43, ALT 115 U/L, AP 358 U/L
- GGT 264
- Amylase 124, Lipase 115
- Na 141, K 4.3, Cl 105, CO_2 26

- Home medication :
  - Omeprazole (20) 1 tab po. od.
  - Cefaclor (250 mg/ml) 5 ml po. tid.
  - Creon (10,000) 1 tab po. tid.
  - CaCO3 (350mg) 1 tab po. od.
  - Ursolin (250 mg) ½ tab po. bid.
- Advice F/U : Familial hypercholesterolemia

Gall stone & bile sludge

- Hemolytic disease
- Hypercholesterolemia
- Anatomical defect
- Drugs
- TPN
- Infection
- Genetic disease: CF
- Idiopathic

F/U OPD

- CBC : Hct 38 %, Hb 13.1 md/dl, WBC 6,900 /mm^3
  (P 23, L 68, M 7, E 2) Plt 222,000/ mm^3
  (MCV 77.2, MCH 26.6, MCHC 34.5, RDW14.9)
- LFT : Alb 4.9 , Glob 2.7, TB 0.65, DB 0.5 , AST 38 , ALT 23 ,
  AP 210
- Amylase 119, Lipase 66

	ผู้ป่วย	บิดา	มารดา
Cholesterol	251	274	232
Triglyceride	101	122	70
C-HDL	36	56	67
C-LDL	176	158	113

CBC บิดา : Hct 41.1 (MCV 95, MCH 31.1, MCHC 32.9, RDW 10.8),
WBC 5,800, Plt 246,000
Blood sugar 90

CBC มารดา : Hct 38.5 (MCV 81, MCH 26.0, MCHC 32.2, RDW 12.1),
WBC 8,400, Plt 177,000
Blood sugar 77

Simon Broome diagnostic criteria for familial hypercholesterolaemia (FH)

Definite

- **TC >6.7 mmol/l (259 mg/dl)** and **LDL >4.0 mmol/l (150mg/dl**), and tendon xanthomas.
- c DNA-based evidence of a LDL receptor mutation, familial defective apo B-100, or a PCSK9 mutation.

Possible **:** cholesterol concentrations as defined above and at least one of the following

- FmHx of myocardial infarction.
- **FmHx** of raised total cholesterol: **>7.5 mmol/l (290mg/dl**) in adult first- or second-degree relative or >6.7 mmol/l in child, brother or sister aged younger than 16 years.

	ผู้ป่วย	บิดา	มารดา
Cholesterol	251	274	232
Triglyceride	101	122	70
C-HDL	36	56	67
C-LDL	176	158	113

Investigation

- CBC : Hct 34.3%, Hb 11.9 md/dl, WBC 11,410 /mm^3(P 74, L 20, M 6)
  Plt 262,000/ mm^3 (MCV 75.1, MCH 26, MCHC 34.7, RDW 15.4)
- LFT :Alb 4.6 , Glob 3.4 g/dL,, TB 4.03 , DB 3.35 mg/dl,
  AST 345 , ALT 647 U/L, AP 678 U/L
- GGT : 707 U/L
- Na 140, K 3.7, Cl 105, CO_2 22
- BUN 10, Creatinine 0.43
- U/A : pH 6.0, sp.gr. 1.015, Protein : negative, Sugar : normal
  Bilirubin 1+, Urobilinogen 1+

- Amylase 565 U/L (0-100)
- Lipase 1,696 U/L (<60)

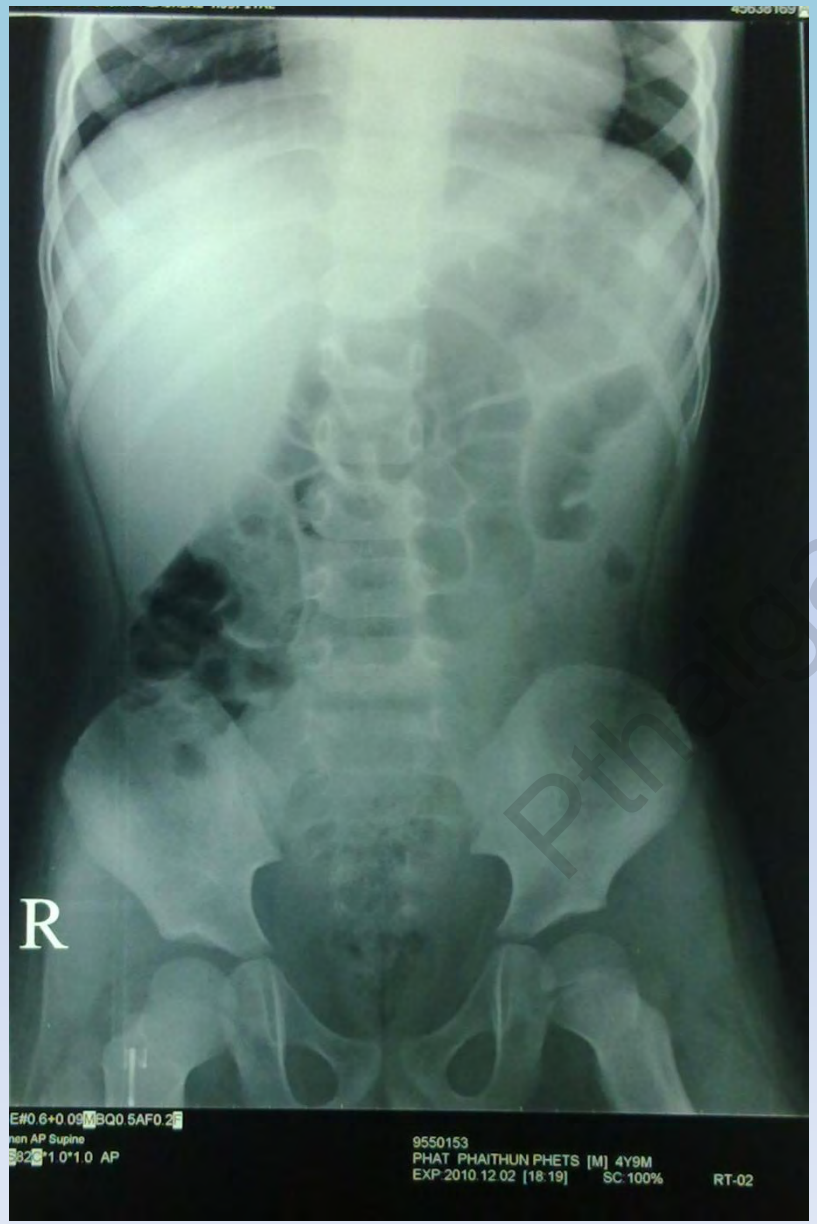
- Total cholesterol : 292 mg/dL
- Triglygeride : 92 mg/dL

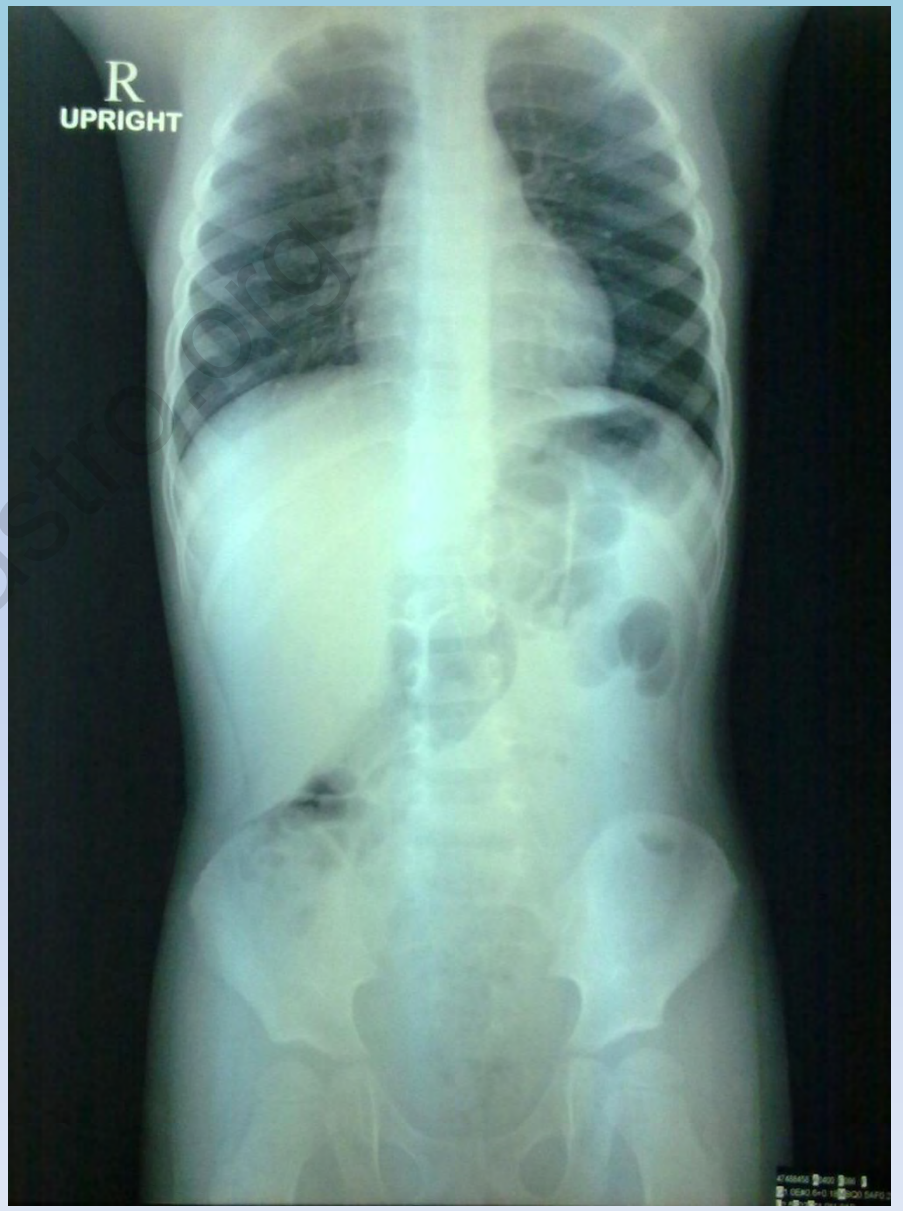
- CMV IgG : negative , IgM : negative
- EBV IgG : Positive , IgM : negative
- HBsAg : negative, Anti-HBc : negative,

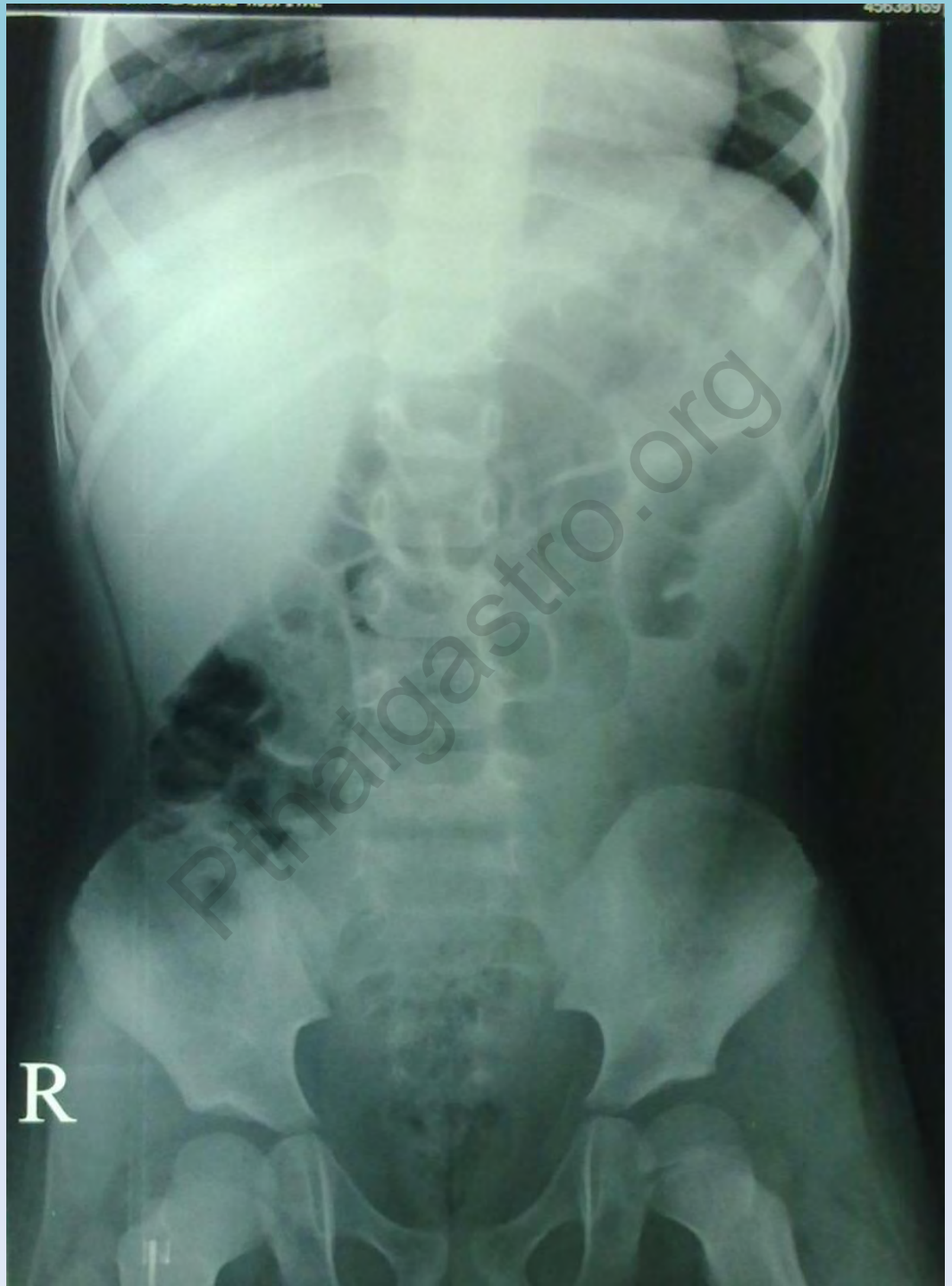
  Anti-HBs : positive (41.6)
- Anti-HAV IgG : negative, IgM : negative
- Anti-HCV : negative

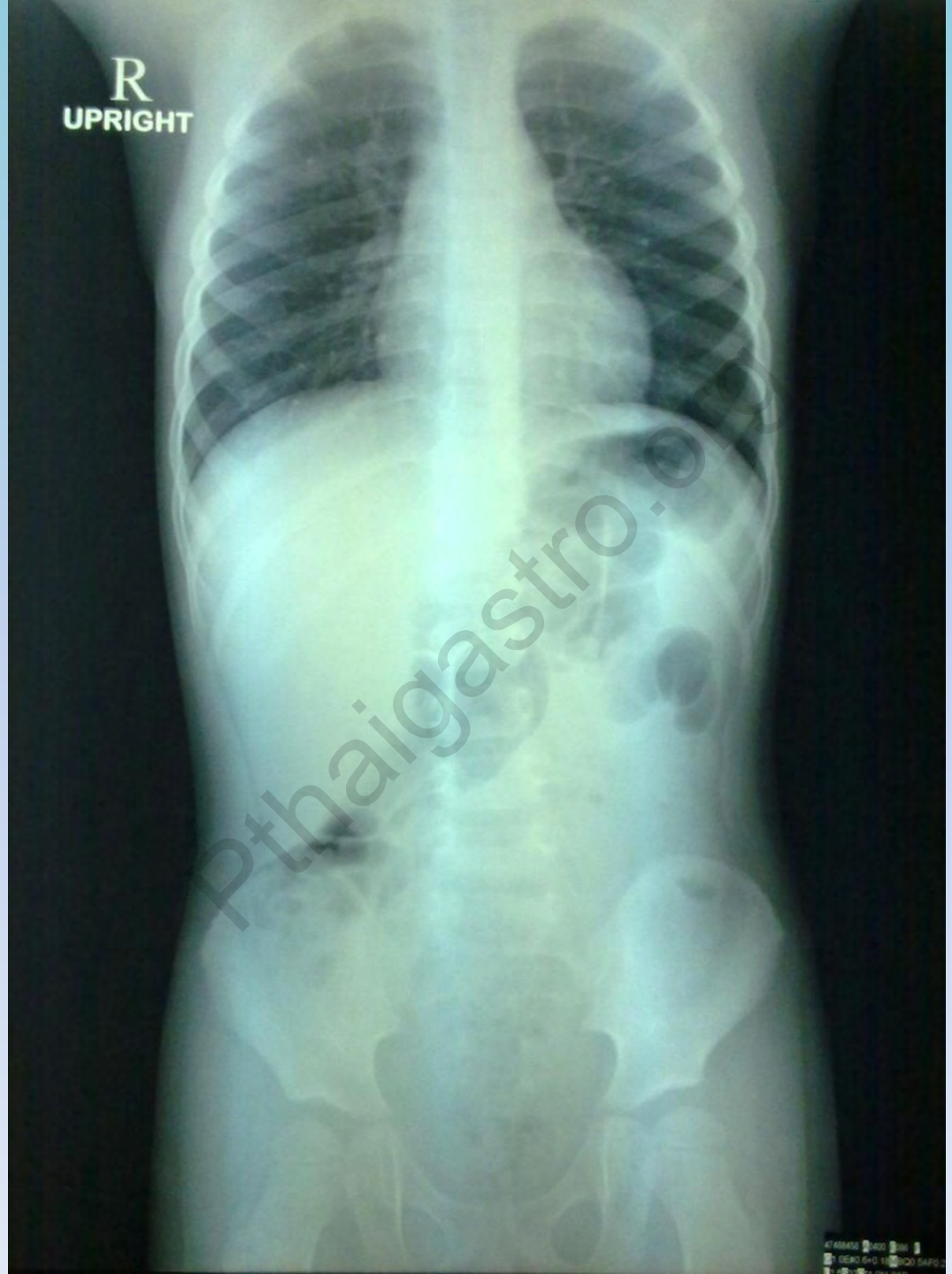
- PT 12.7 (12.2), PTT 27.6 (27), INR 1.04
- Blood sugar 96
- CRP < 3.41 mg/L (<5)
- ESR : 28 mm/hr
- Hemoculture : no growth
- Hemoglobin typing : Normal

Film abdomen

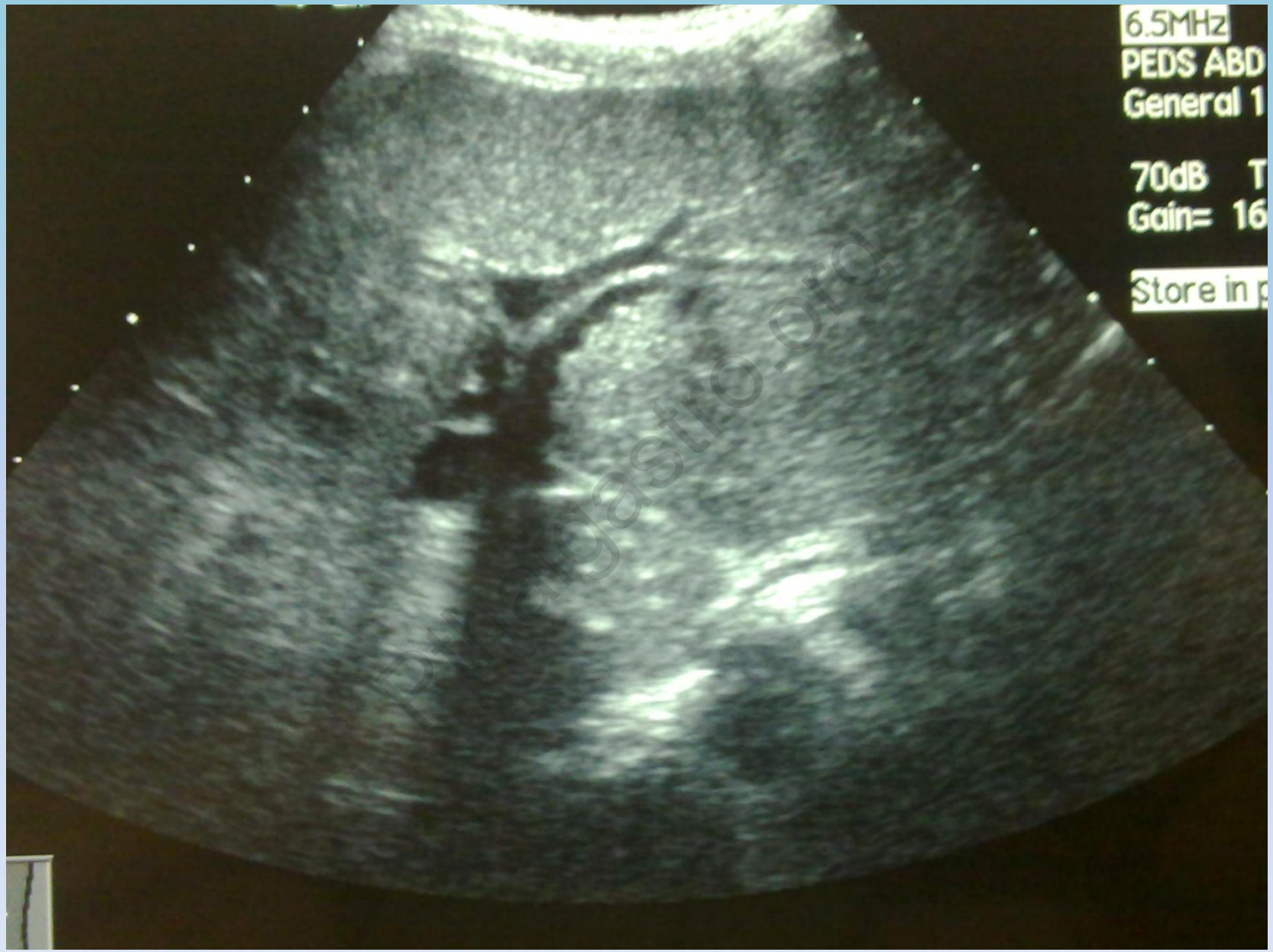


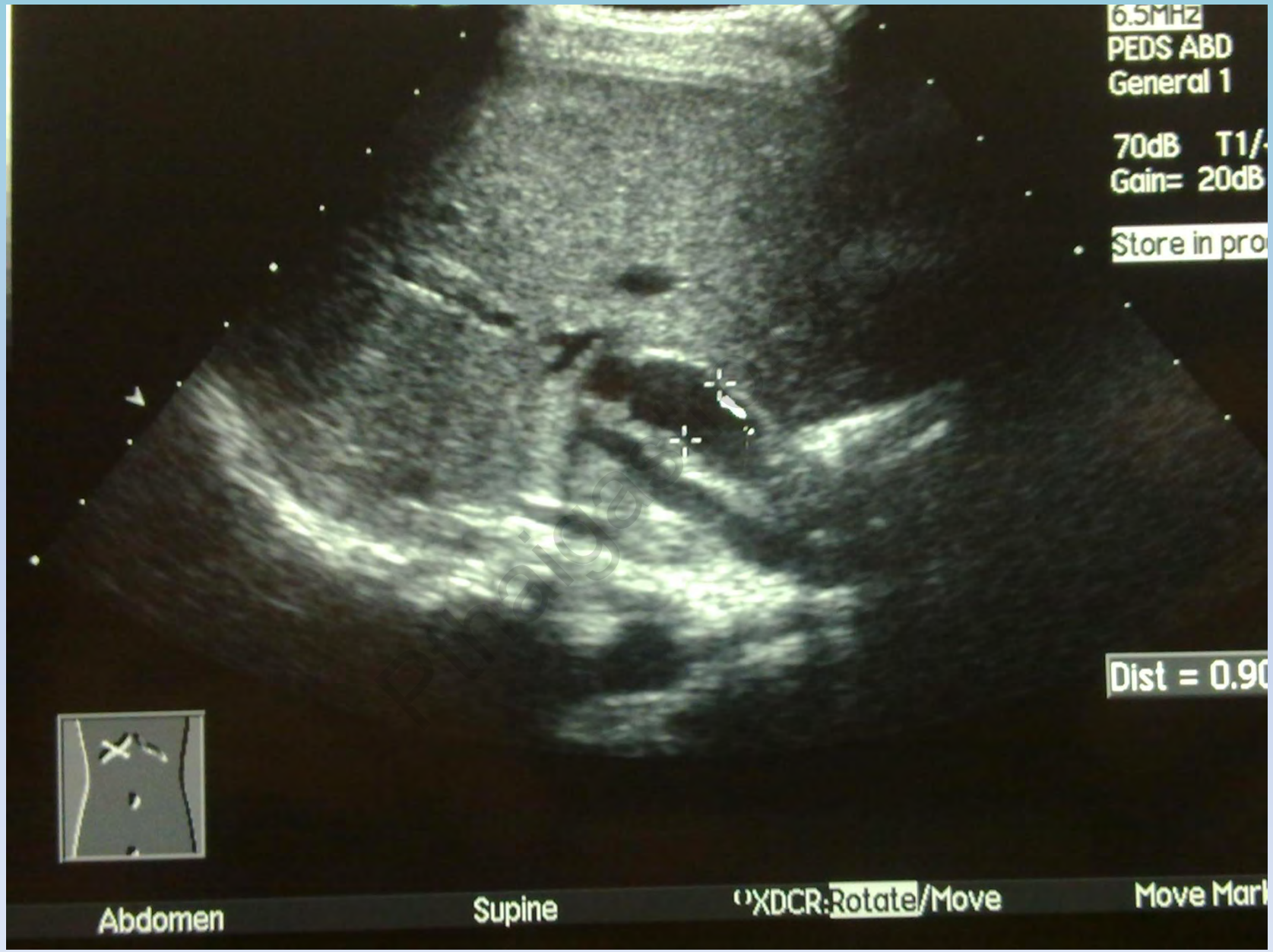


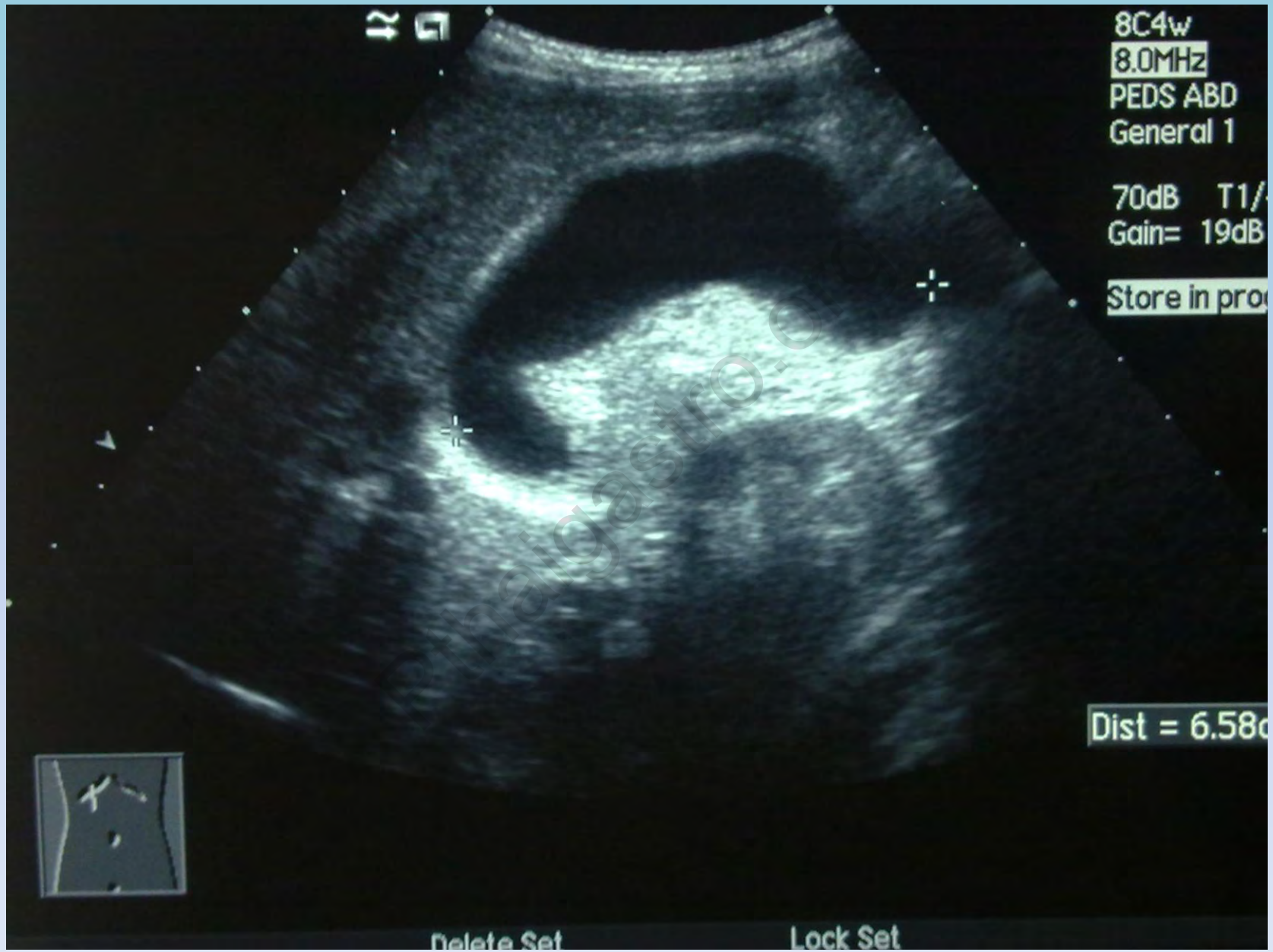


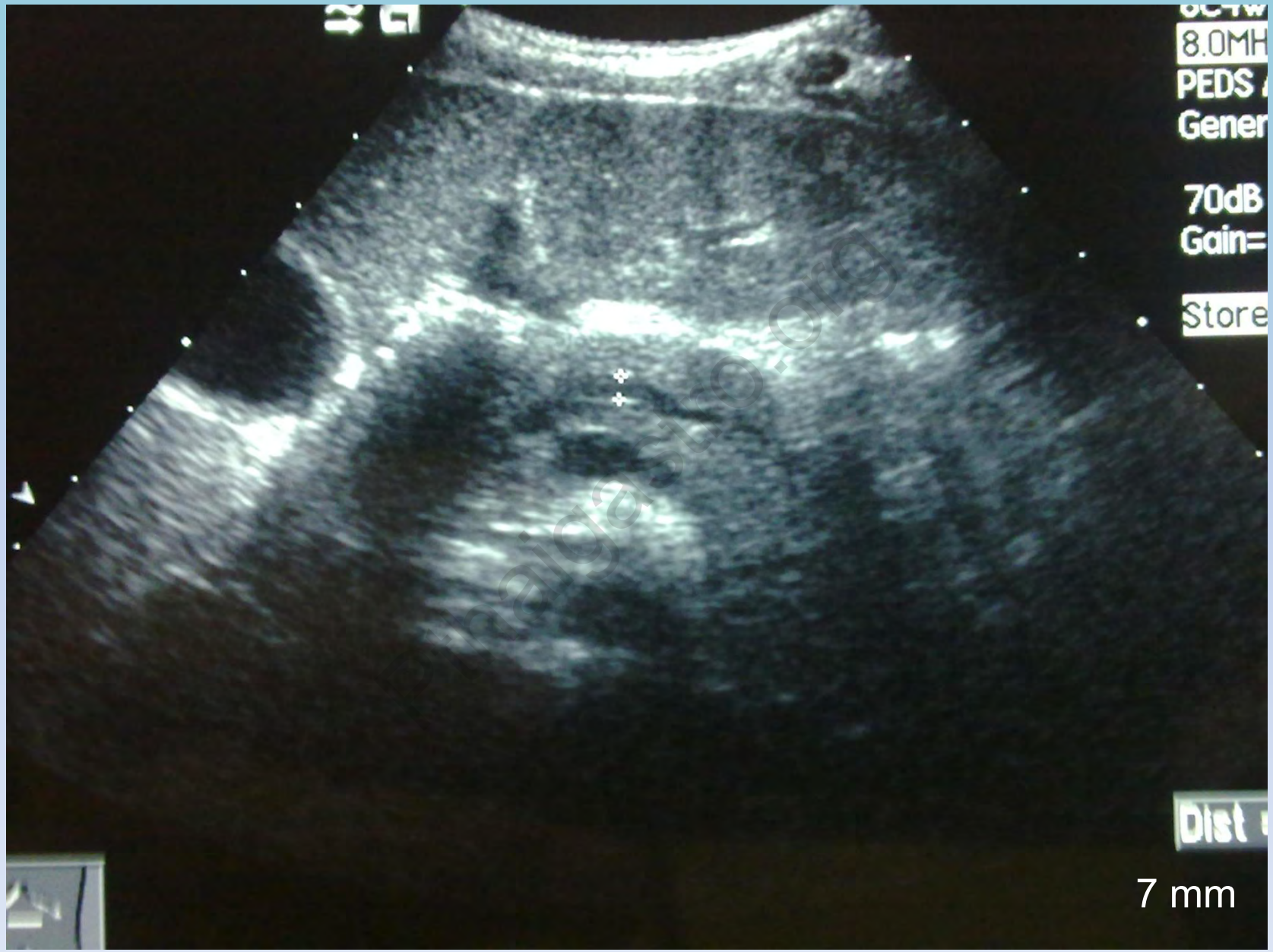


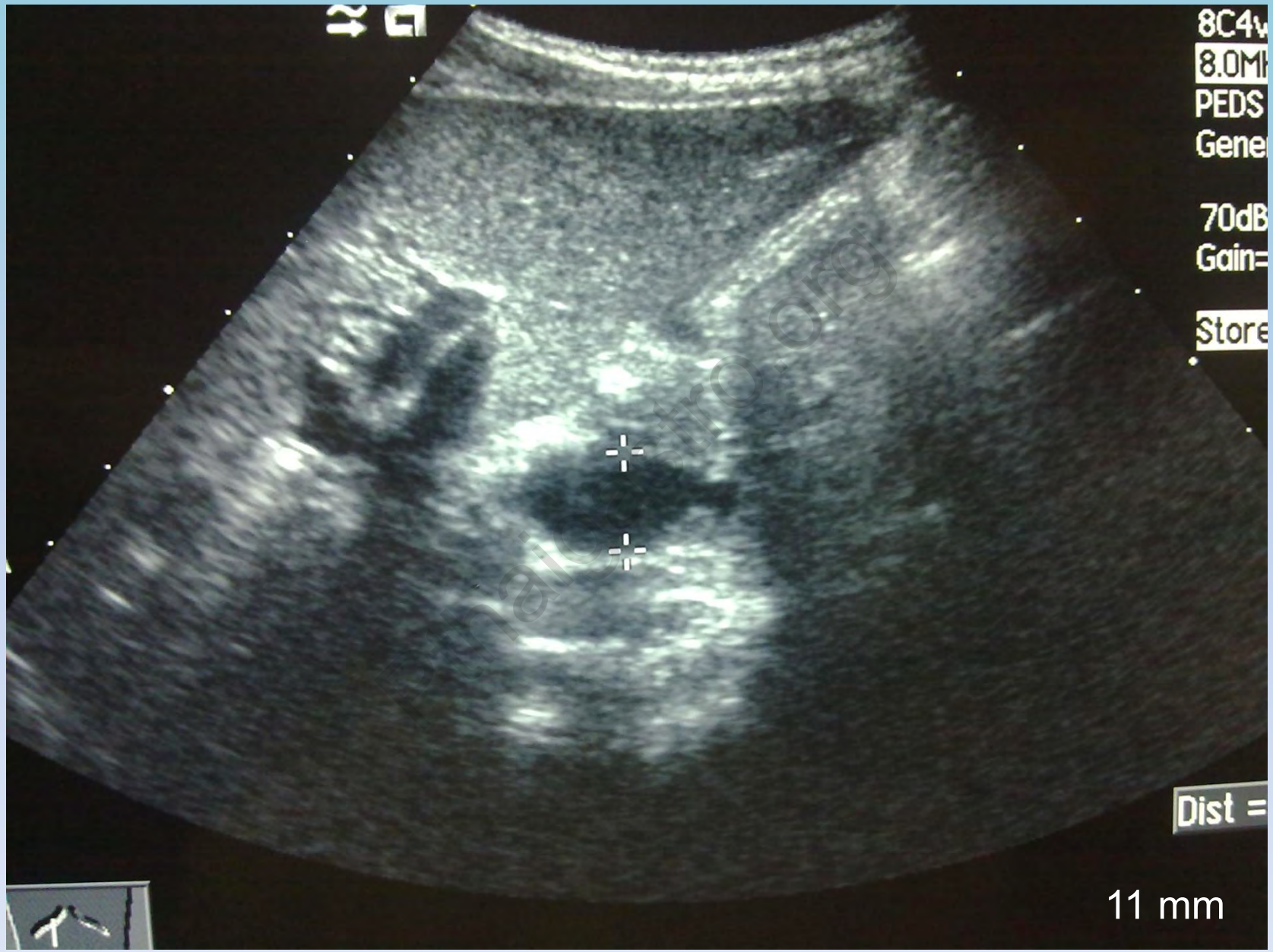
Ultrasound upper abdomen

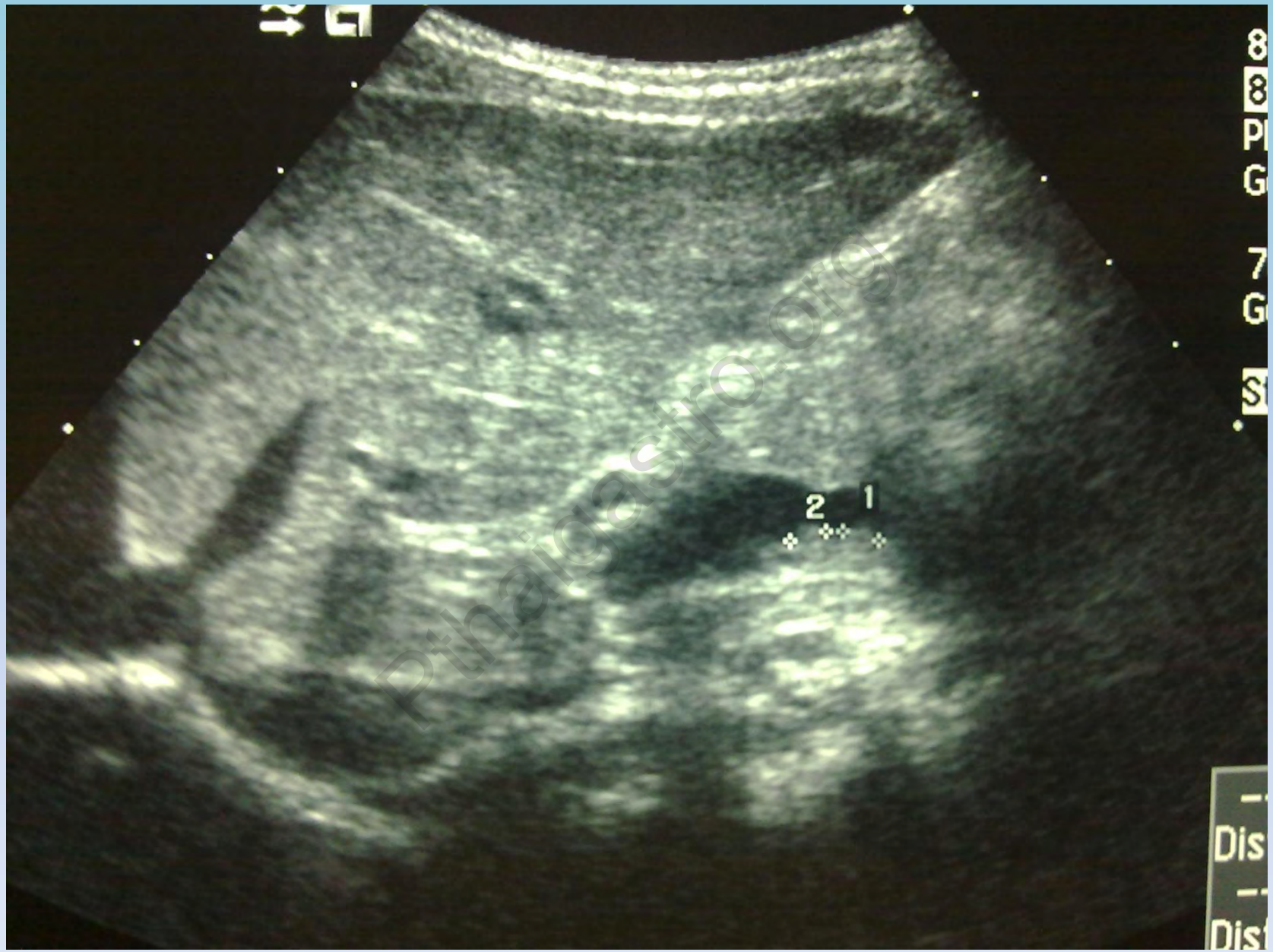

6.5MHz
PEDS ABD
General 1
70dB T
Gain= 16
Store in
Pregastric.org

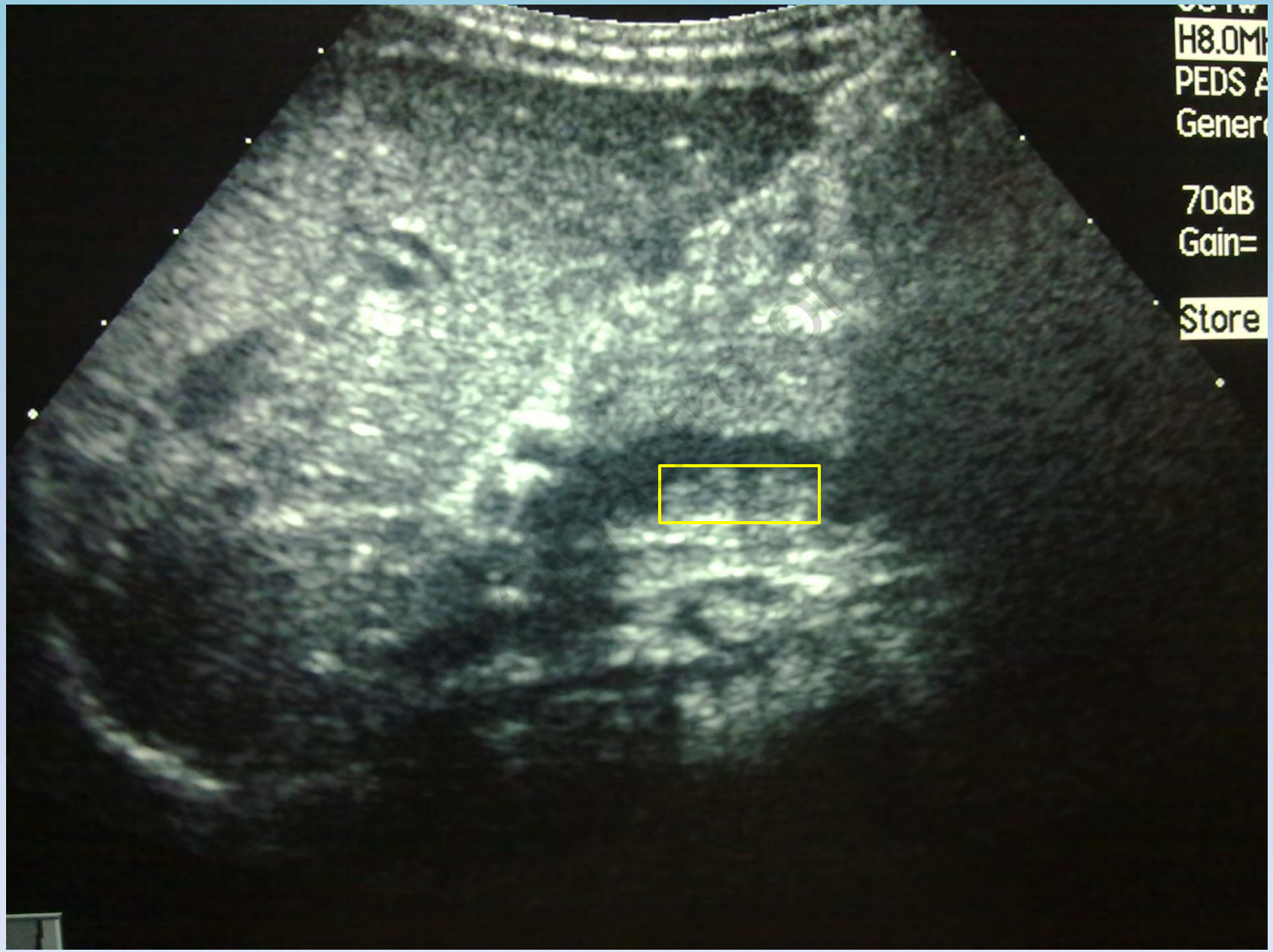

6.5MHz
PEDS ABD
General 1
70dB T1/
Gain= 20dB
Store in pro
Dist = 0.90
Abdomen
Supine
XDCR:Rotate/Move
Move Mark


8C4w
8.0MHz
PEDS ABD
General 1
70dB
Gain= 19dB
Store in pro
Dist = 6.58
Delete Set
Lock Set


8.0MH
PEDS
Gener
70dB
Gain=
Store
Dist
7 mm


8C4v
8.0MH
PEDS
Gene
70dB
Gain=
Store
Dist =
11 mm

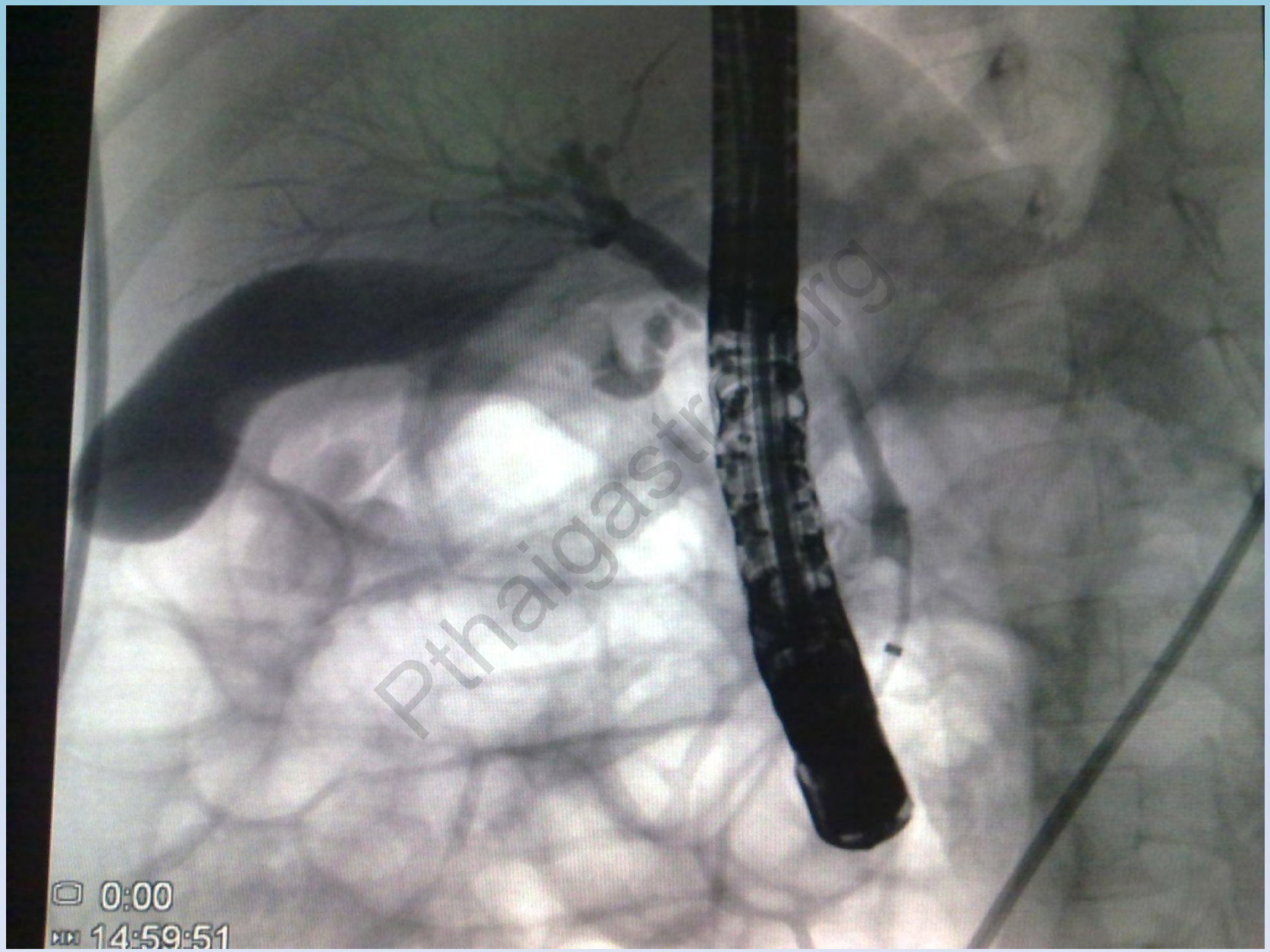

2
1
Pinaigastro.org

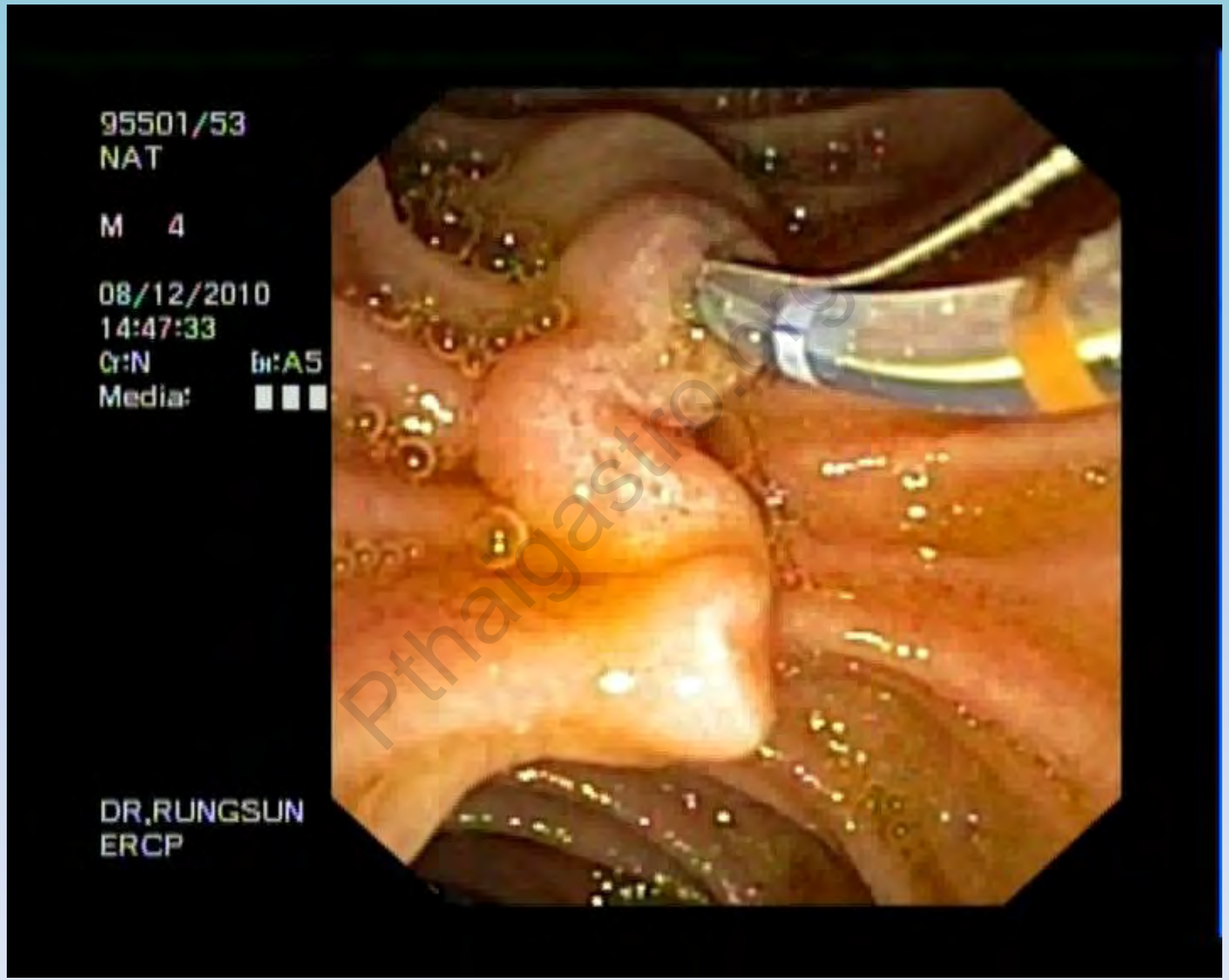

H8.0MH
PEDS A
Generc
70dB
Gain=
Store

Ultrasound upper abdomen

- Liver : normal size, shape and echogenicity, no SOL.
- Diffuse mild dilatation of bilateral IHD
- Moderate dilatation of CBD to distal part (CBD 1.1 cm)
- a few round medium echogenicity small lesions without acoustic shadow within distal CBD (size about 0.4 cm each)

ERCP


0:00
14:59:51
Pthaigastro.org


95501/53
NAT
M 4
08/12/2010
14:47:33
Cr:N
Bi:A5
Media:
DR,RUNGSUN
ERCP
Pthaigastro.org

ERCP :

- **Cholangiogram**

  CBD : CBD sludge, CBD 3 mm in diameter

  Common hepatic duct : normal

  Intrahepatic duct : normal

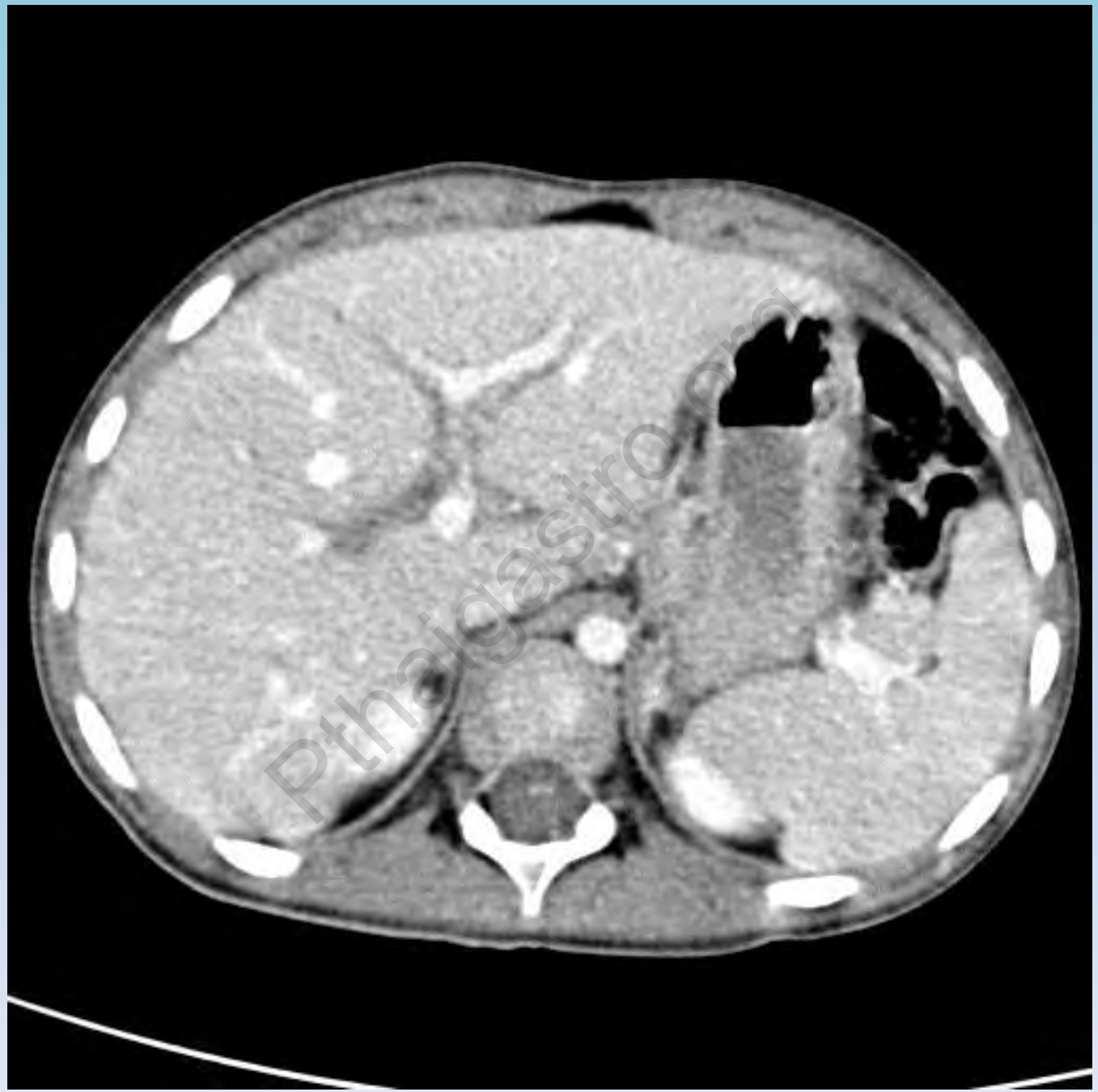
  Rt & Lt intrahepatic duct : normal

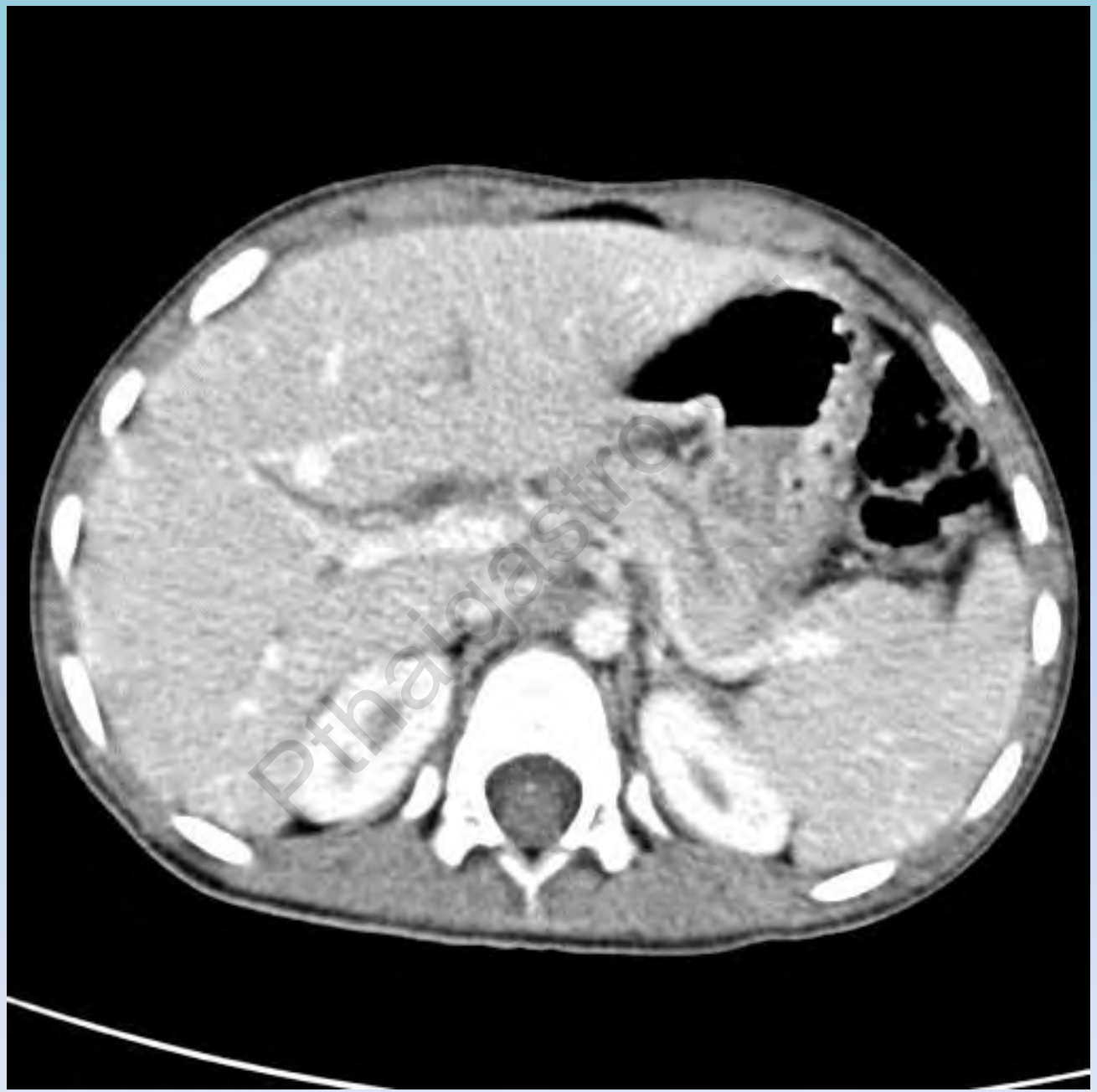
  Cystic duct : normal

- Procedure : standard sphincterotomy, stone extractoin with balloon

  (Balloon extraction reveal bile sludge)

CT upper abdomen


Pthalgastro.org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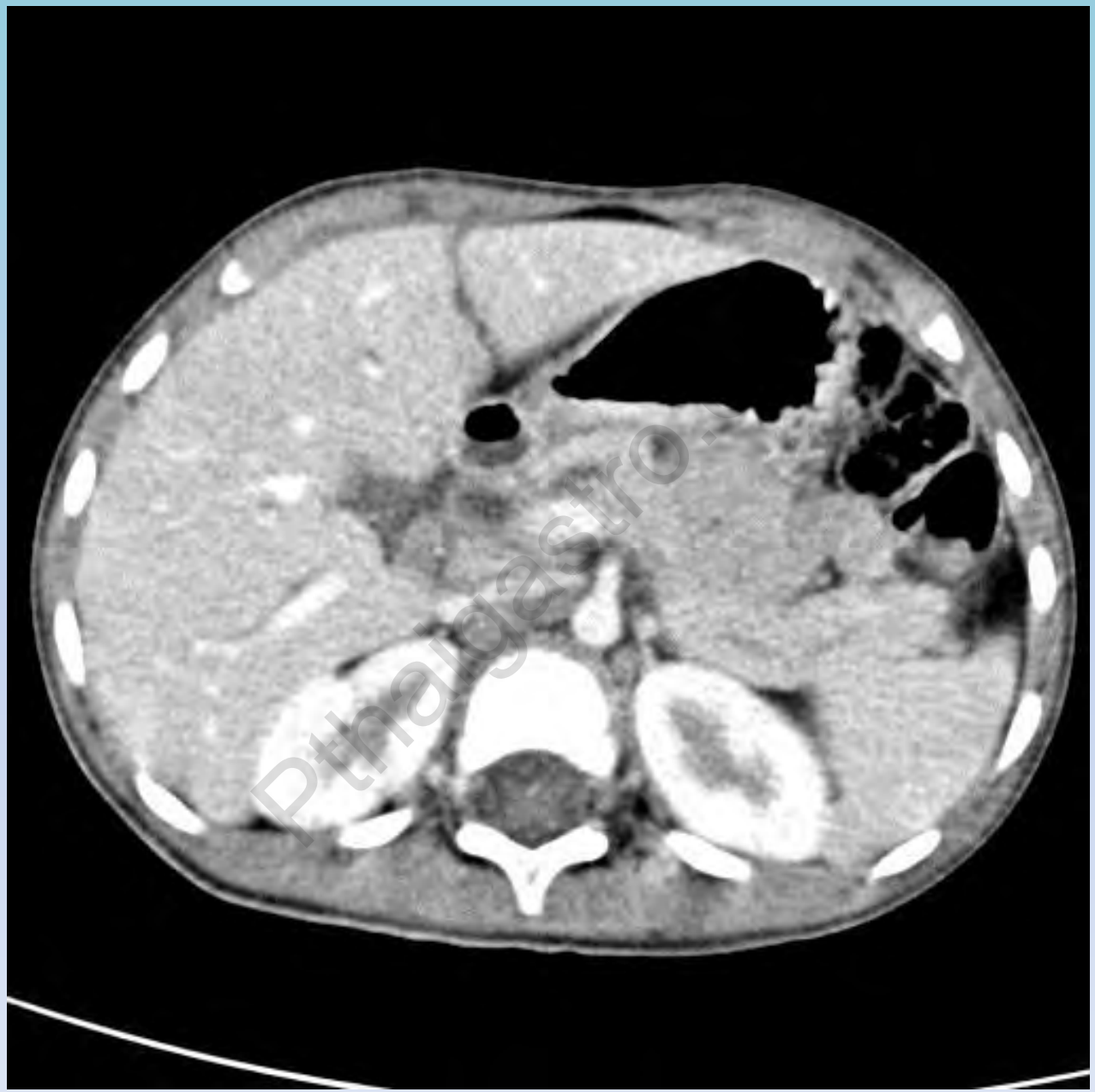
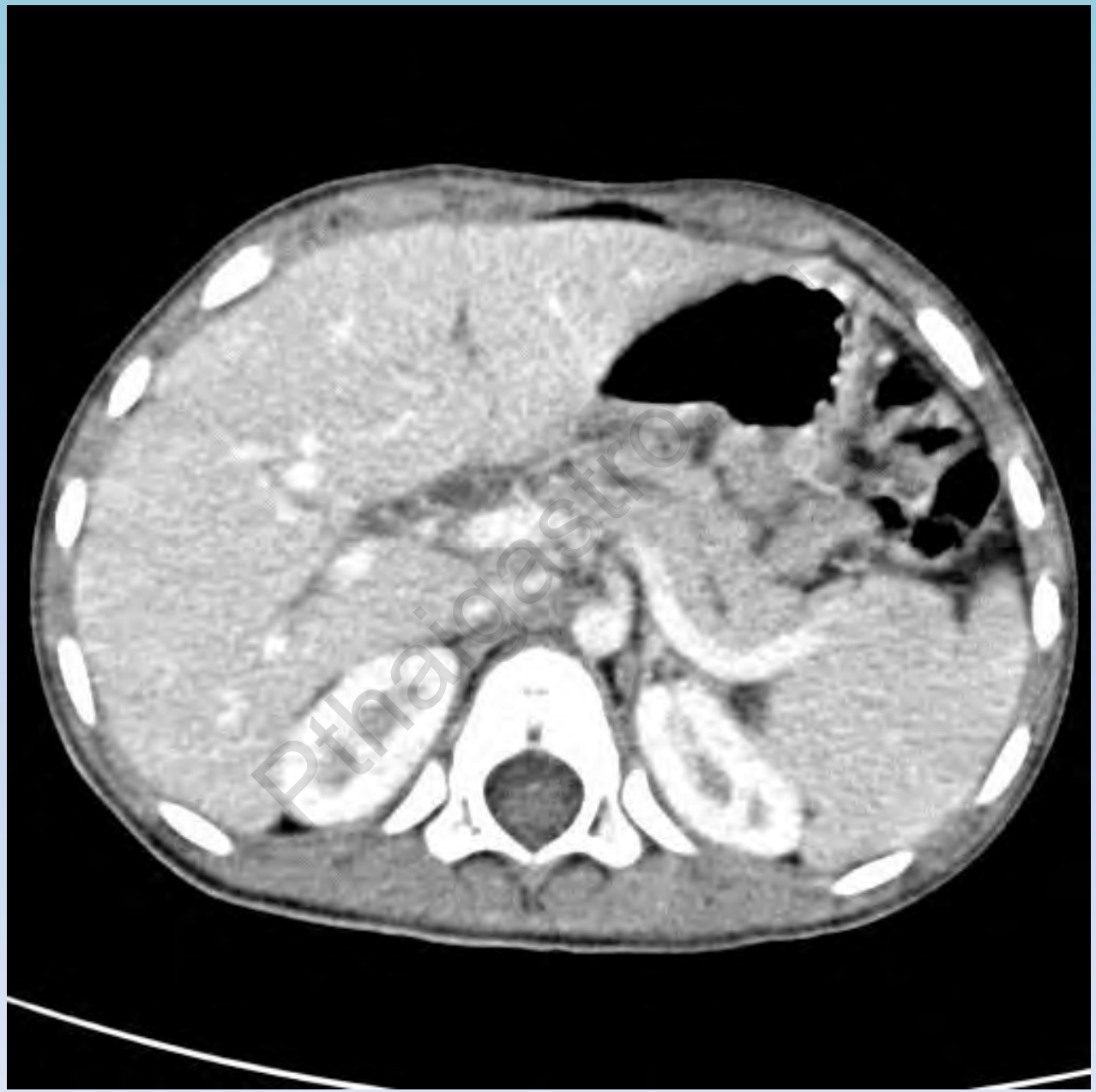

Pthaidastro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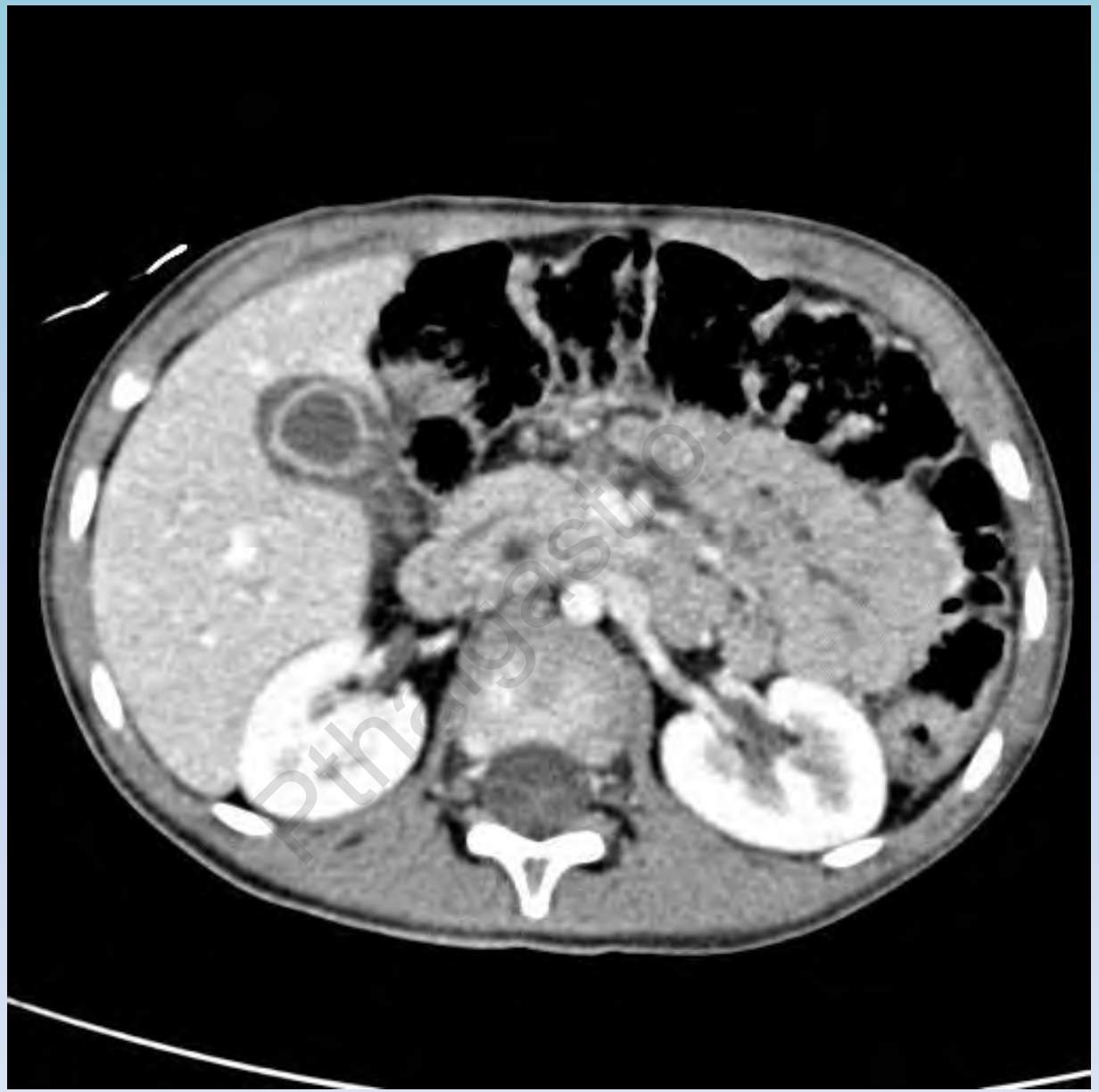
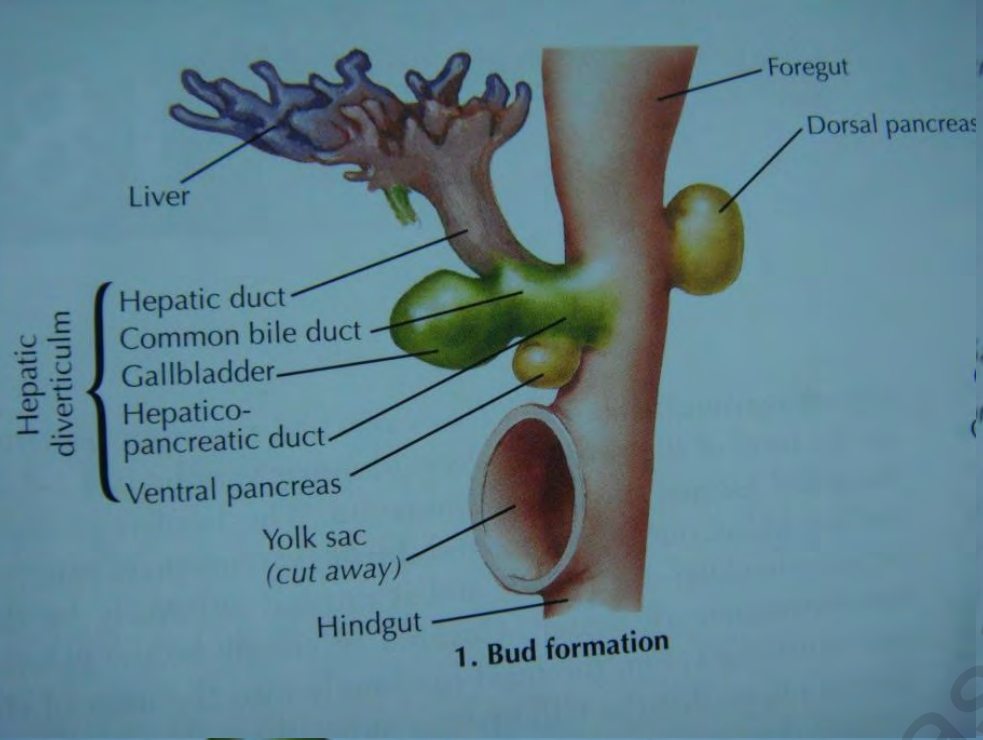

Pthangasuo

CT upper abdomen

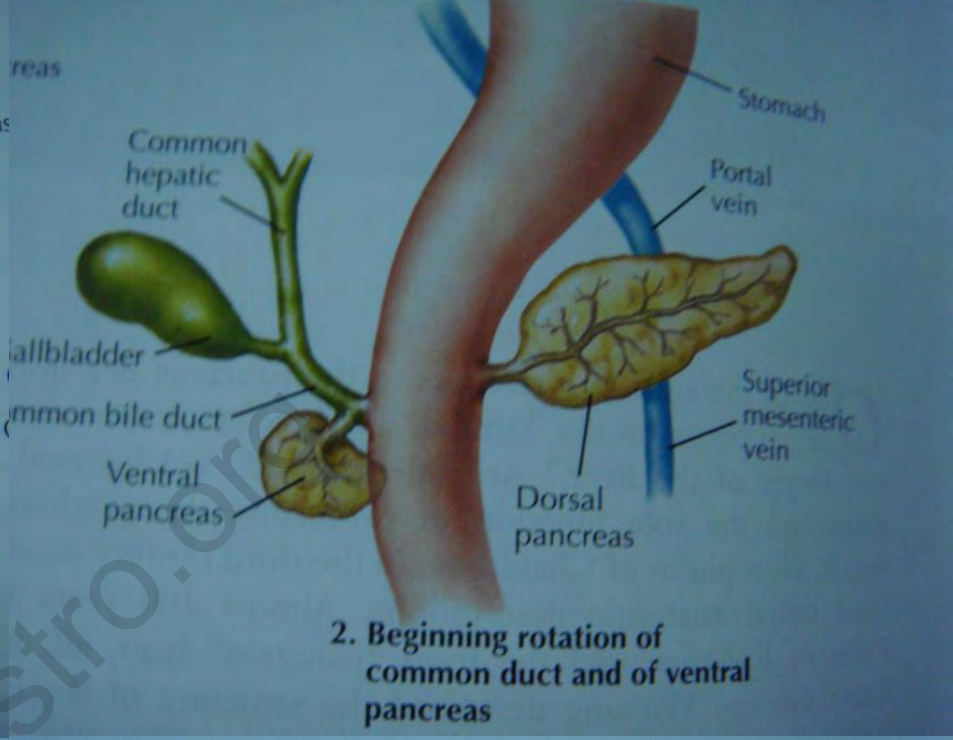
- Liver : normal size, shape and attenuation
- Mild dilatation of bilateral proximal IHD.
- Mild dilatation of CBD (6 mm) down to distal portion and dilatation of main pancreatic duct (3 mm) without opaque intraluminal lesion or extrinsic mass.
- Thickened wall of distal CBD.
- Variation of persistent duct of Santorini with communication with main pancreatic duct.

CT upper abdomen

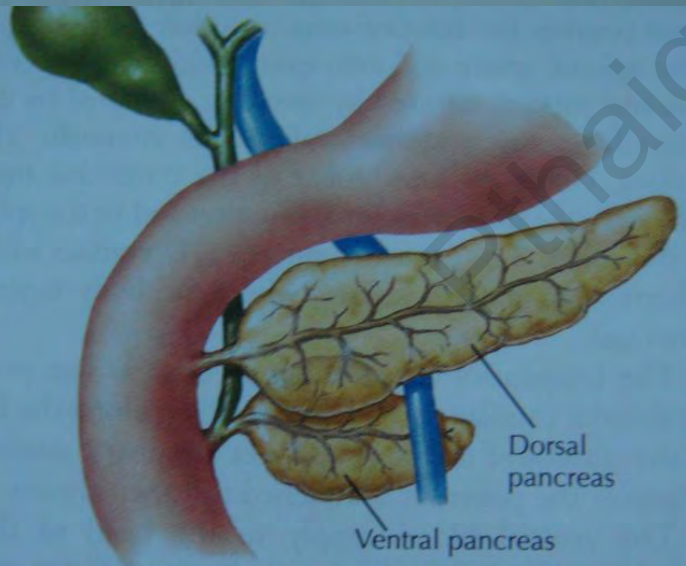
- GB fairly distened, containing no mass and stone.
- Low density of GB fossa, suggestive of edema.
- Pancreas normal size, no mass and calcification.
- Adrenal gland, Both side of kidneys : normal
- No ascites



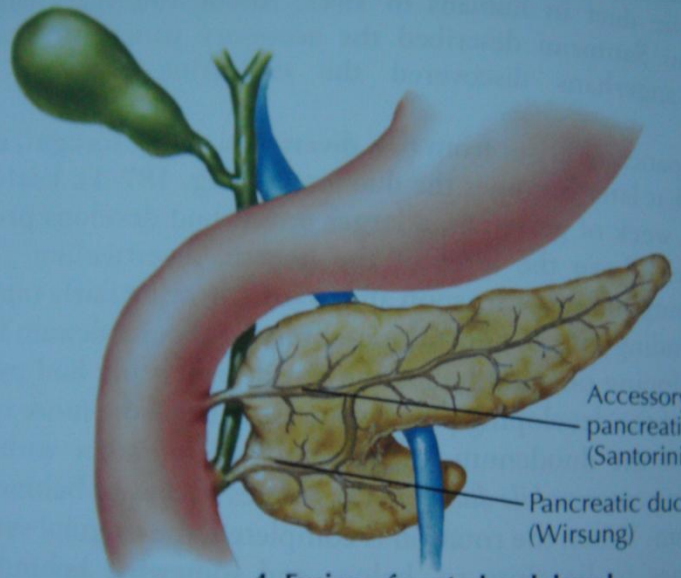
1. Bud formation



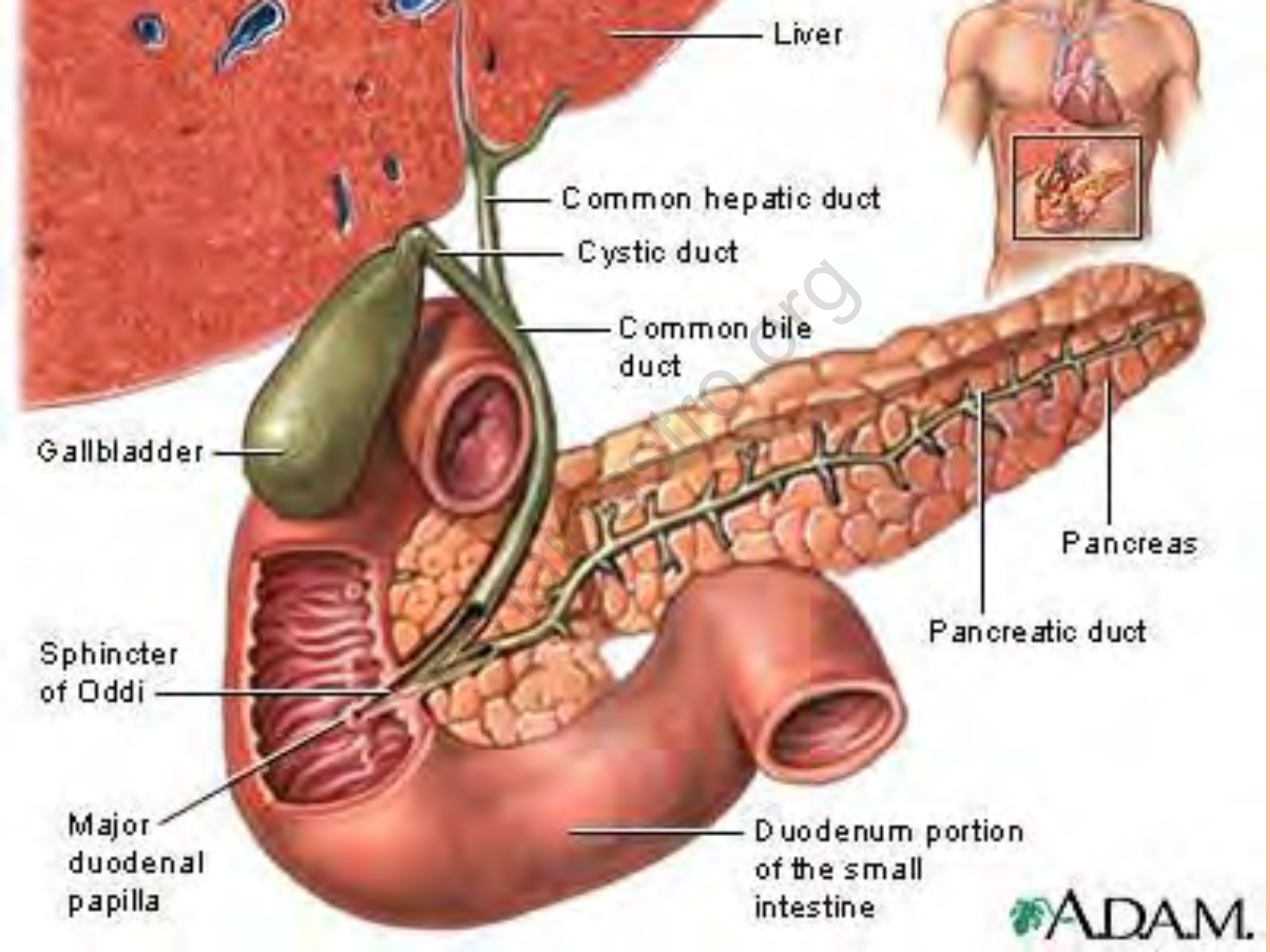
2. Beginning rotation of common duct and of ventral pancreas

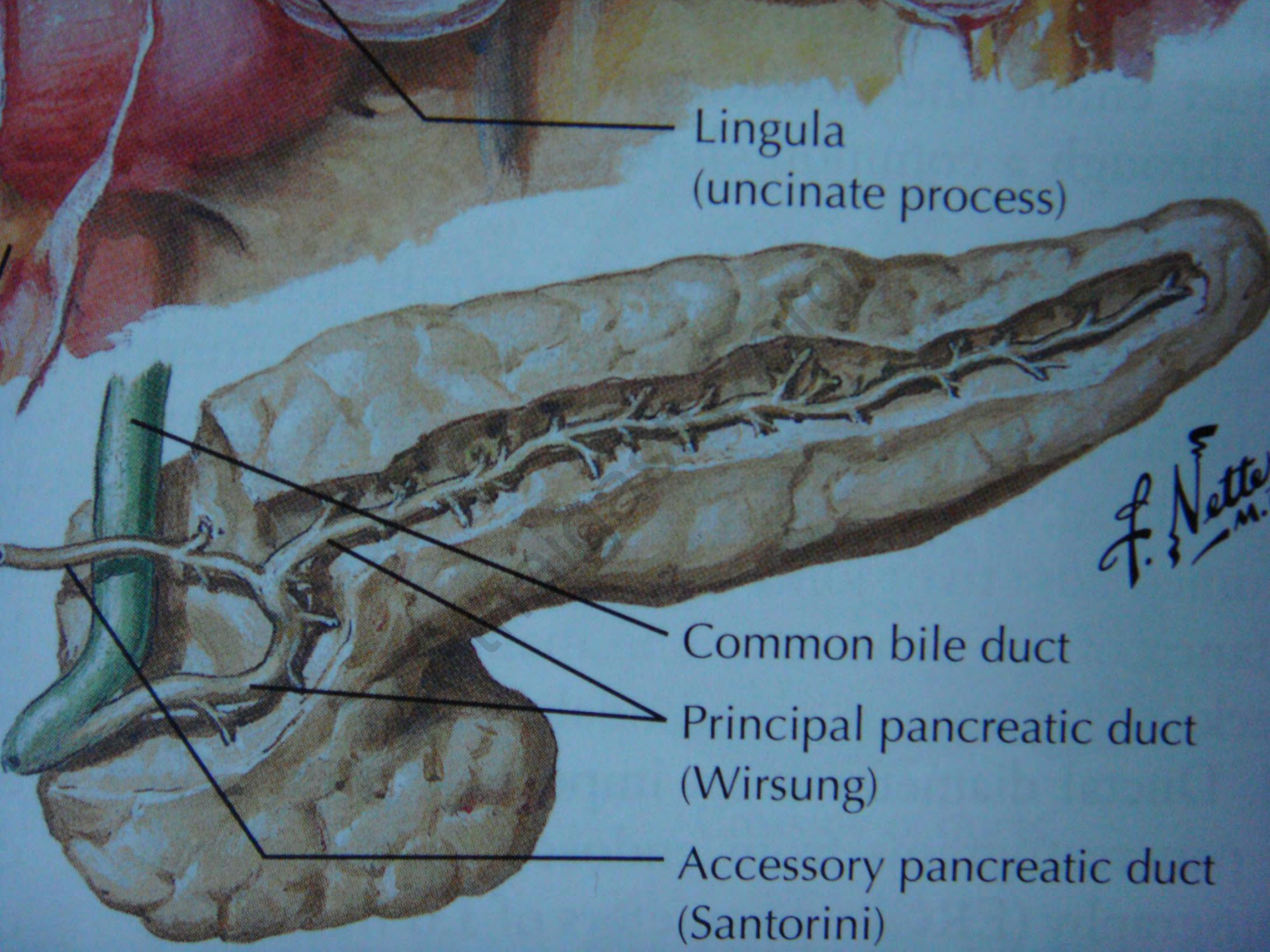


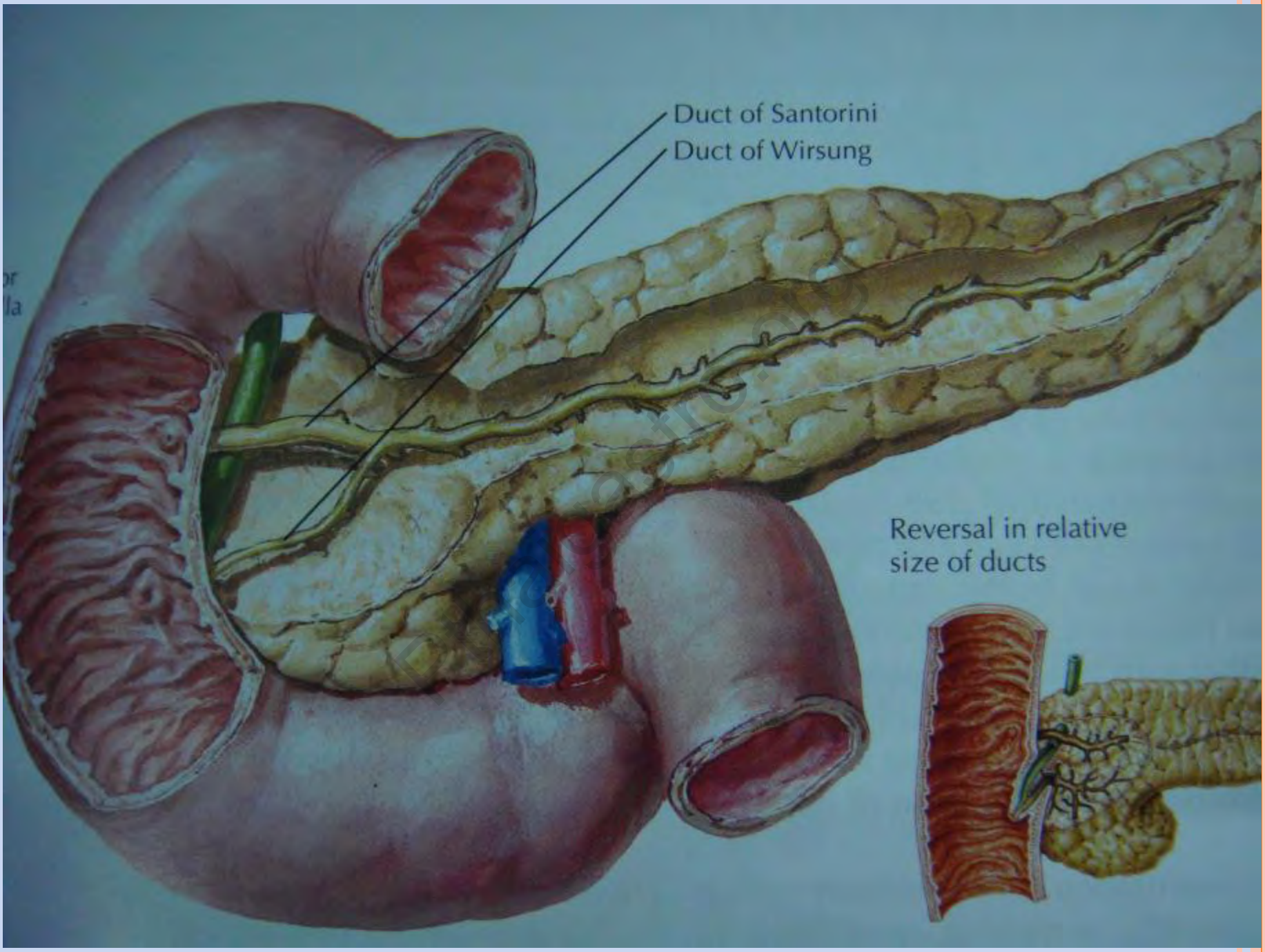
3. Rotation completed but fusion has not yet taken place

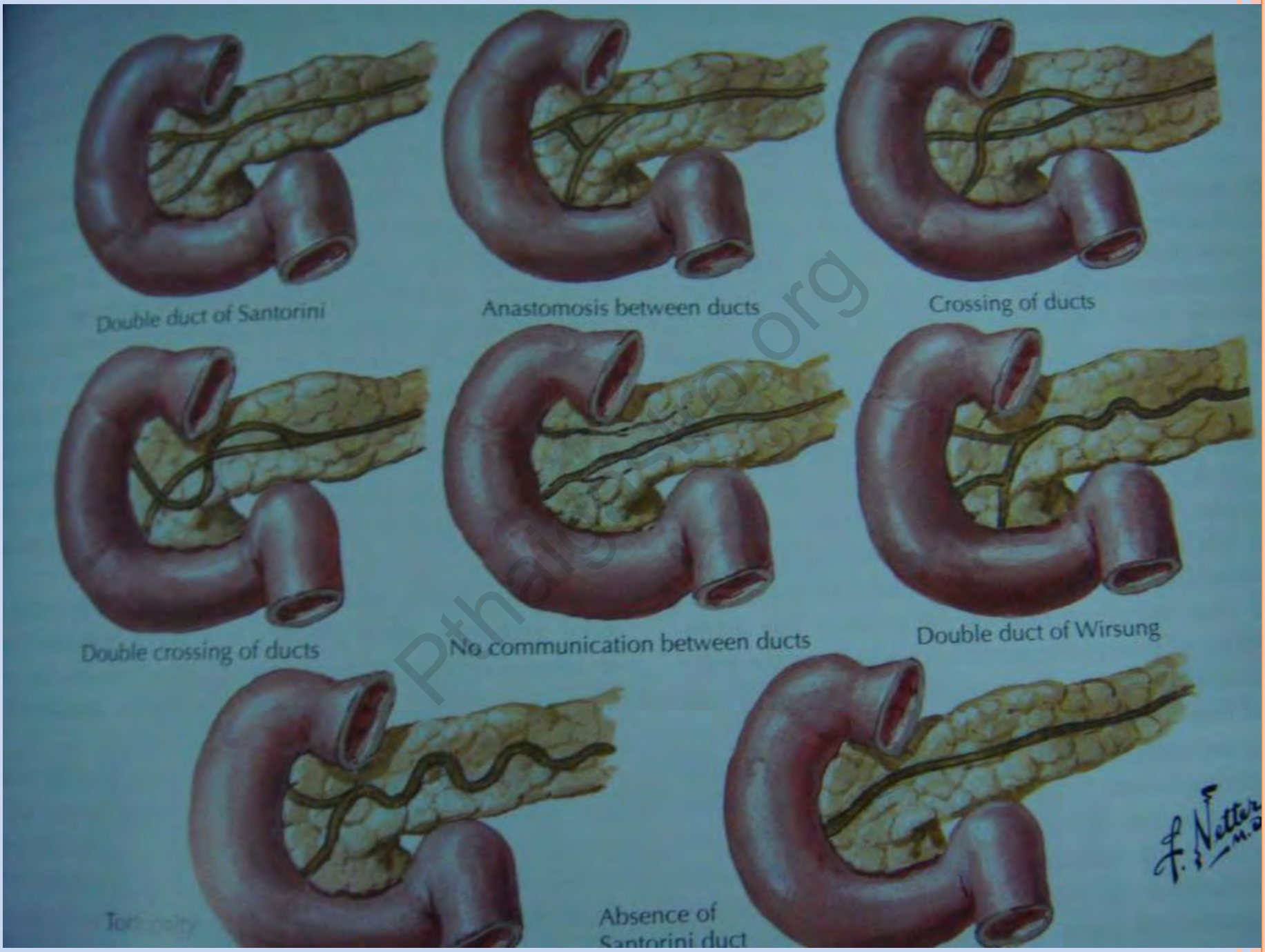


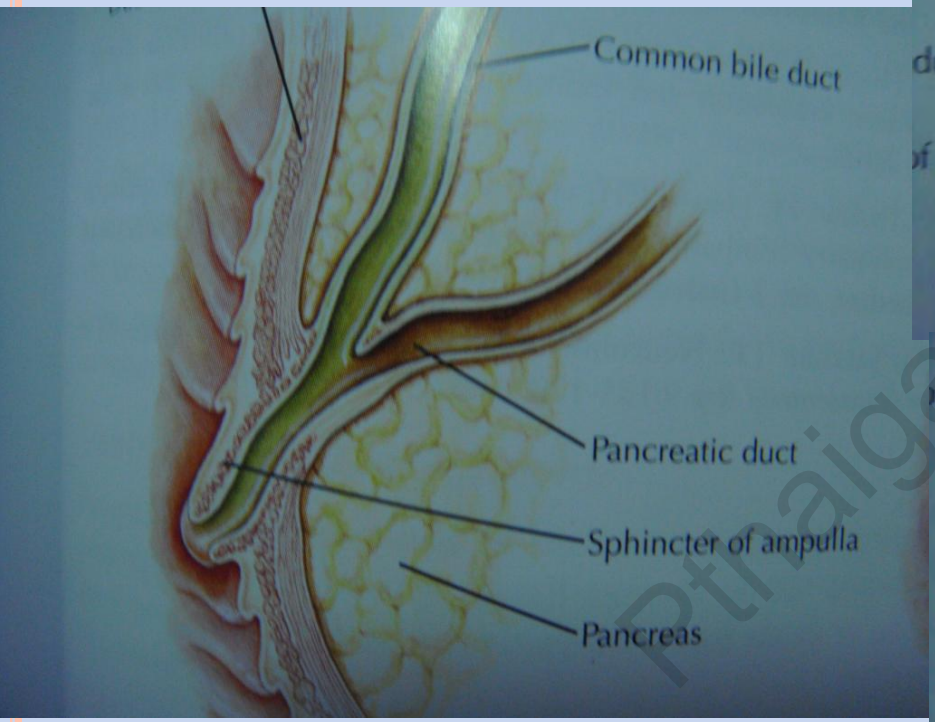
4. Fusion of ventral and dorsal pancreas and union of ducts

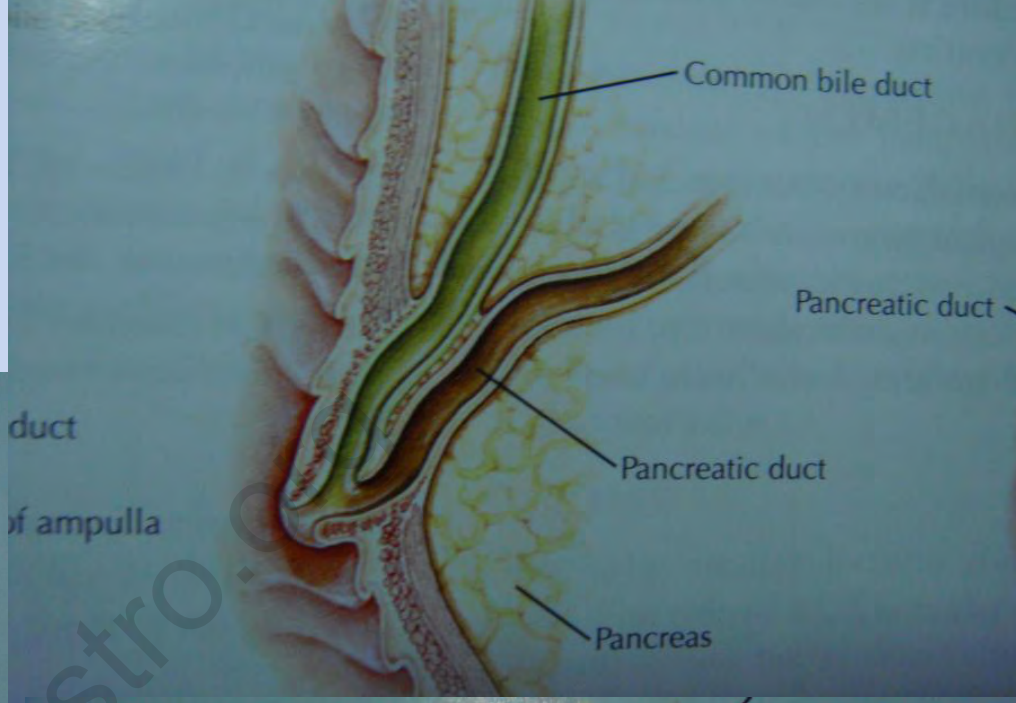

Liver
Common hepatic duct
Cystic duct
Common bile duct
Gallbladder
Pancreas
Pancreatic duct
Sphincter of Oddi
Major duodenal papilla
Duodenum portion of the small intestine
A.D.A.M.

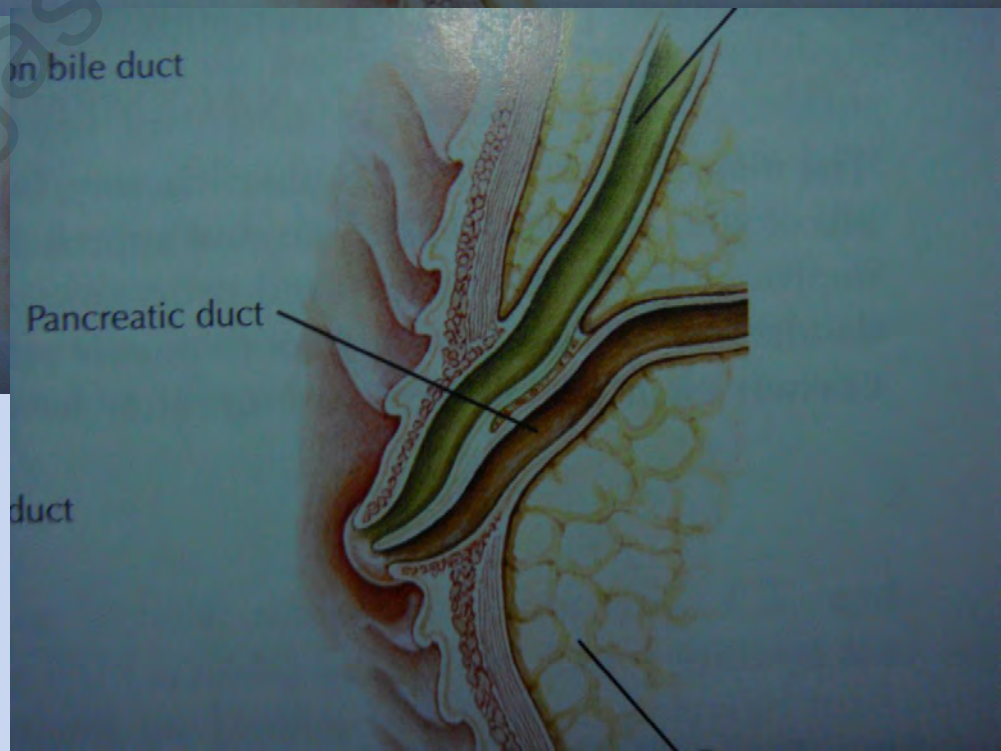

Lingula
(uncinate process)
Common bile duct
Principal pancreatic duct
(Wirsung)
Accessory pancreatic duct
(Santorini)
F. Netter M.D.


Duct of Santorini
Duct of Wirsung
Reversal in relative size of ducts

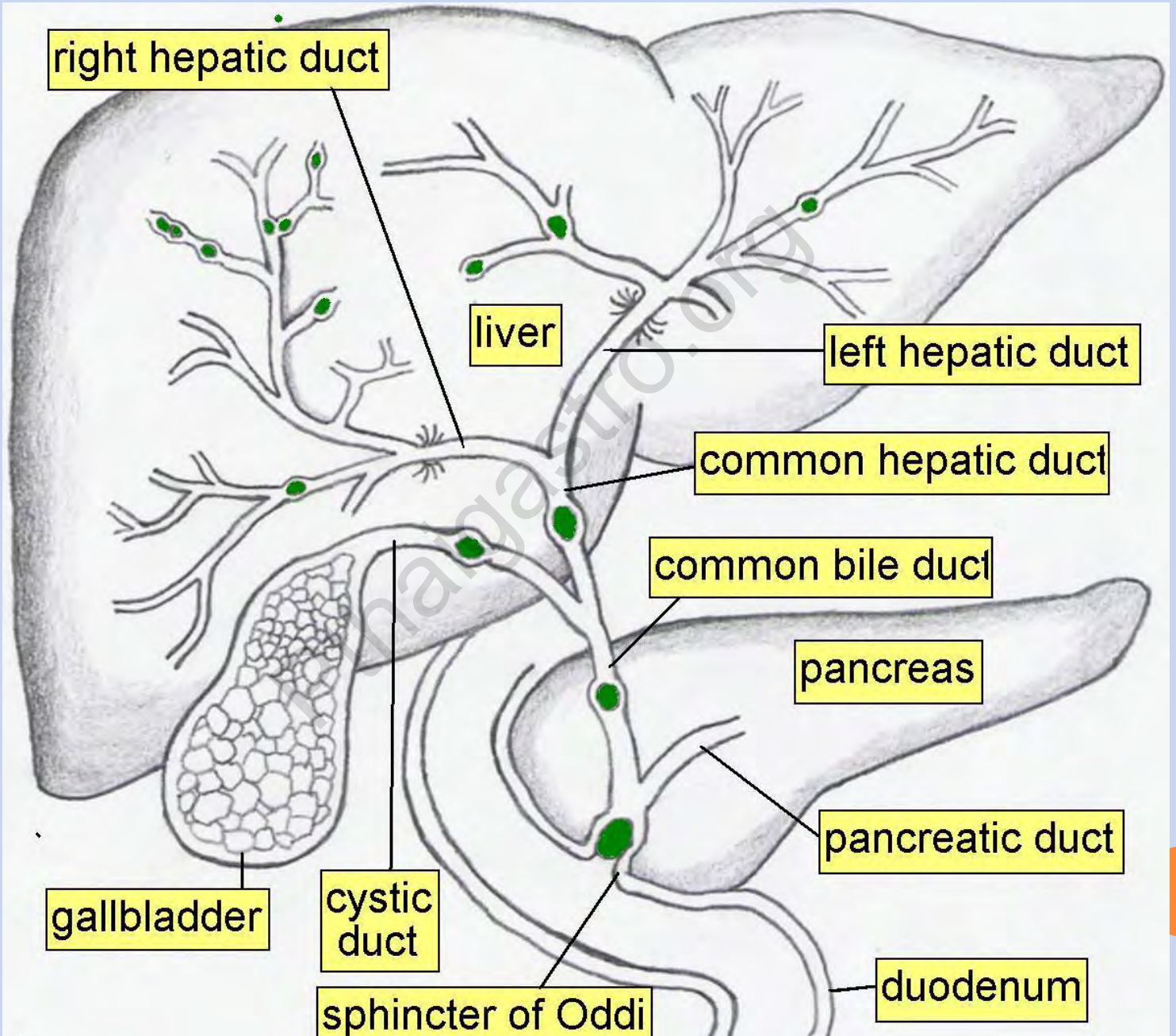

Double duct of Santorini
Anastomosis between ducts
Crossing of ducts
Double crossing of ducts
No communication between ducts
Double duct of Wirsung
Absence of

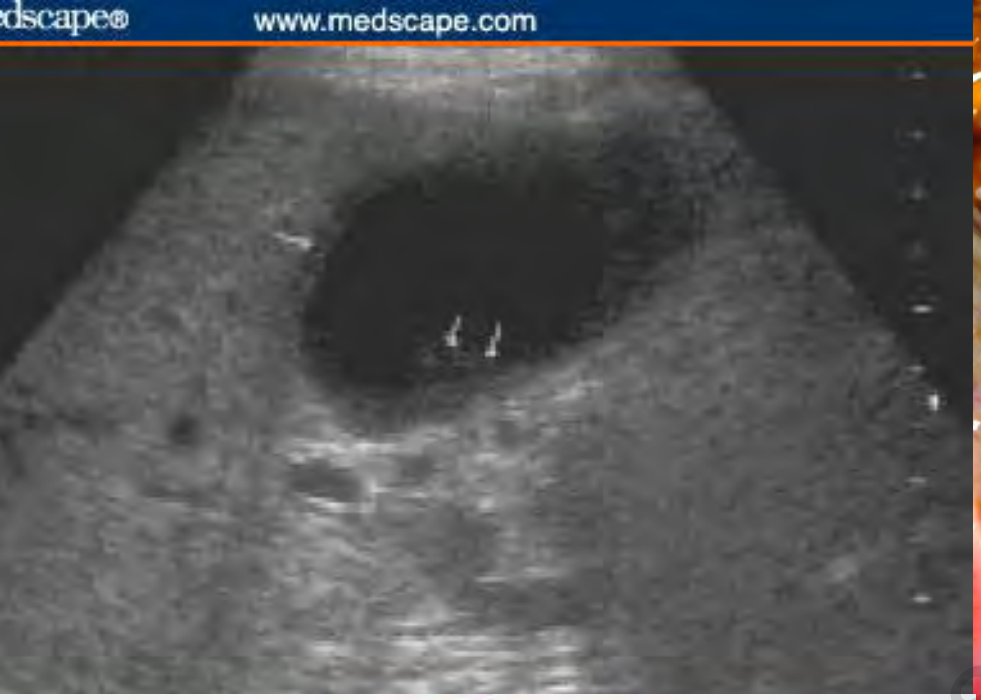






GALLSTONE, SLUDGE

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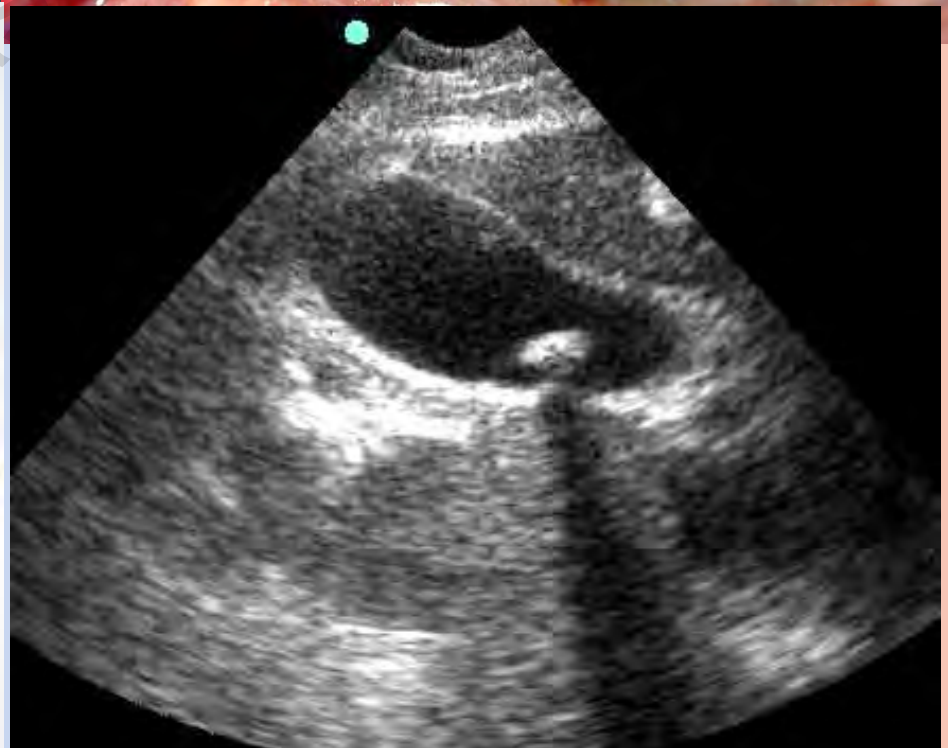


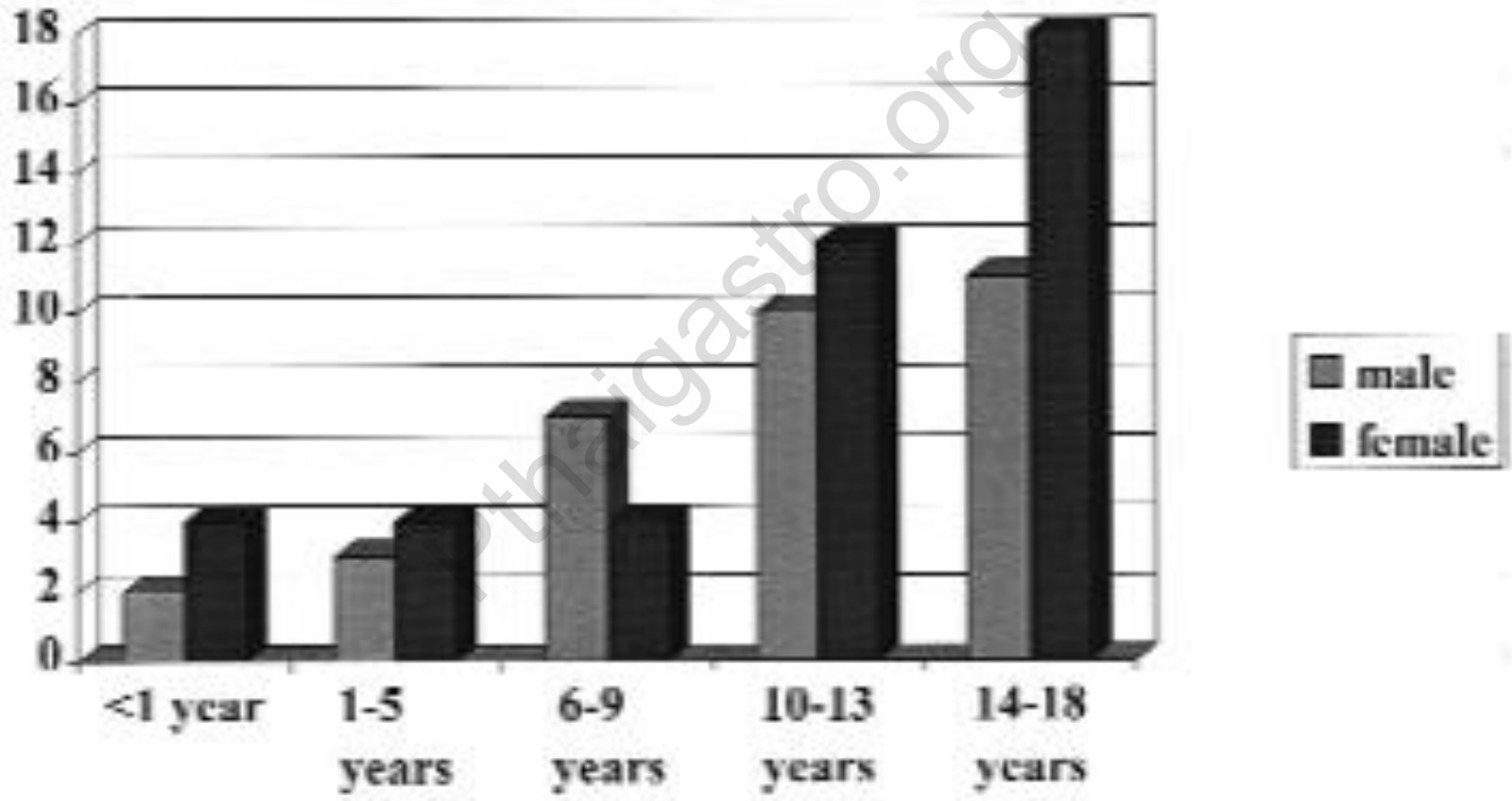
Type	Location	Composition	Risk factor
Cholesterol	Gallbladder	Cholesterol monohydrate	Female, age, diet, rapid weight loss, drug, obesity, dyslipidemia
Black pigment	Biliary tract + gall bladder	Calcium bilirubinate	Hemolysis
Brown pigment	Intrahepatic duct	Calcium carbonate + fatty acid	Infection
Biliary sludge	Variable	Variable	Prolong fasting, TPN, drugs (ceftriaxone)

David Q., et al. Gallstone disease, 1089-1189

Cholelithiasis and biliary sludge in children

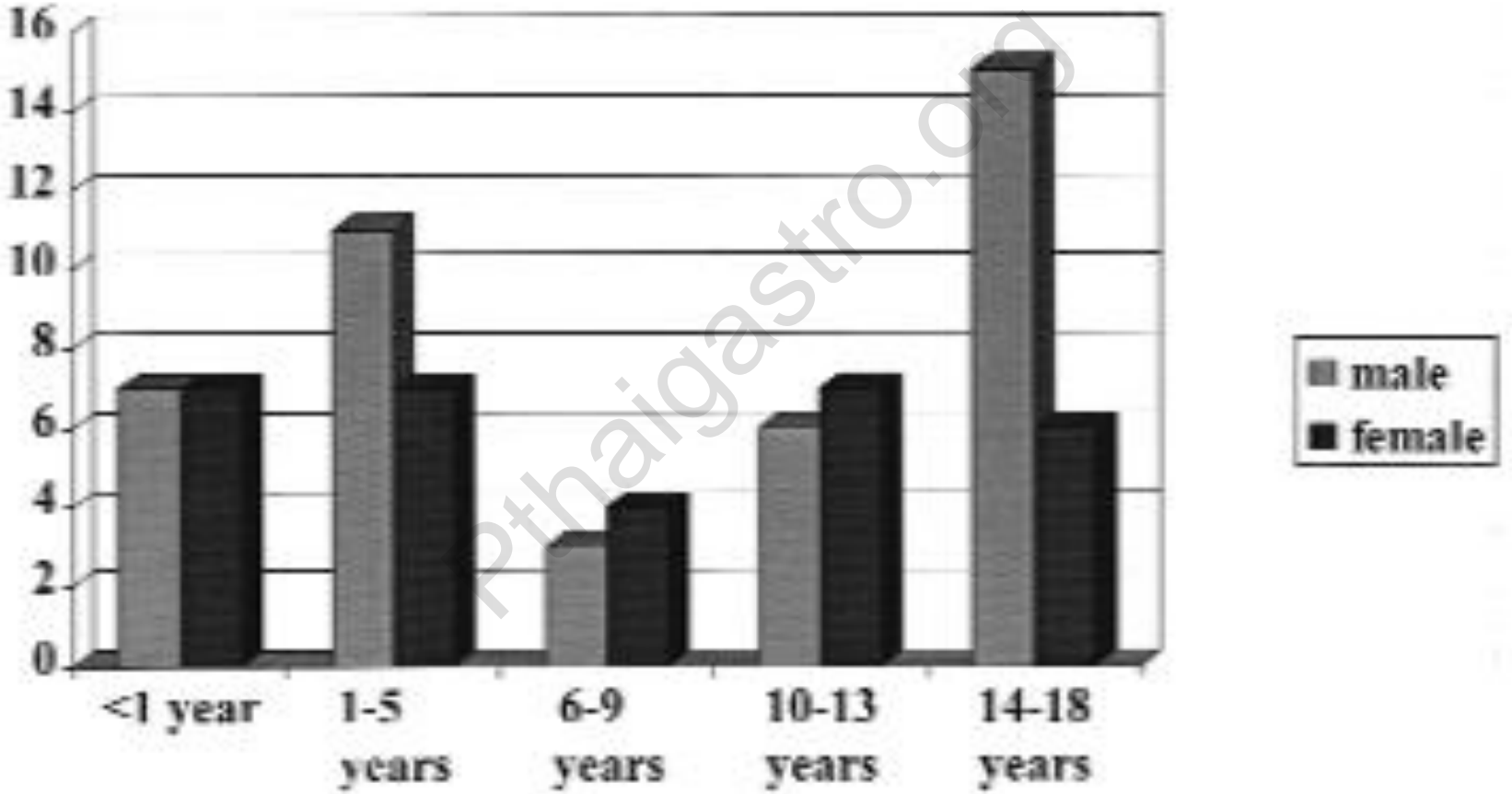
Isebel W, et al. JPGN 2000. 31:411-417

Age distribution pattern with cholelithiasis



Isebel W, et al. JPGN 2000. 31:411-417

Age distribution pattern with biliary sludge



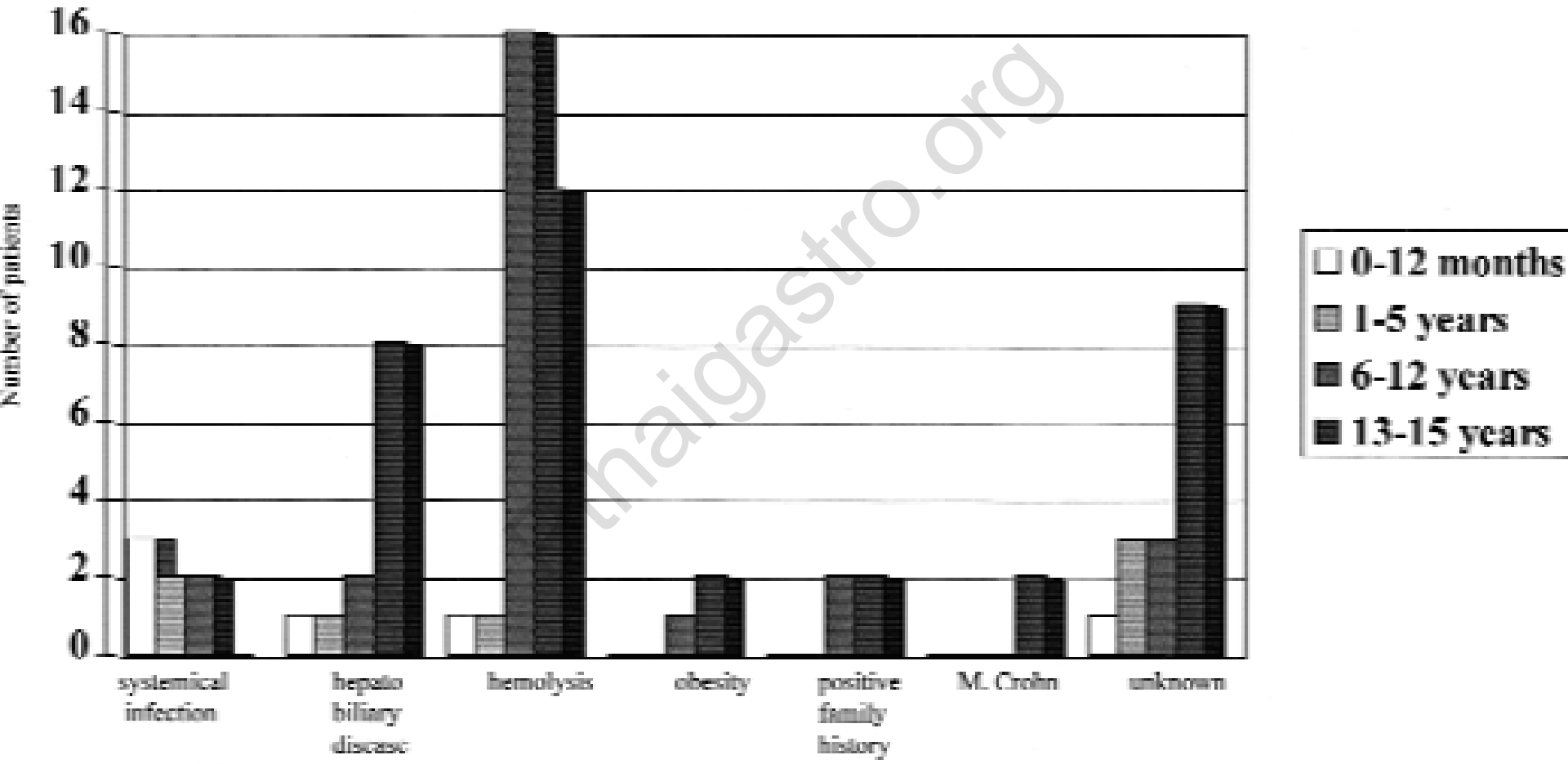
Isebel W, et al. JPGN 2000. 31:411-417

Association condition with gallstones and sludge

Associated condition	Patients with gallstones (n)	Patients with sludge (n)
Hemolytic disease	32	5
Hepatobiliary disease	13	10
Systemic infection or antibiotic use	6	22
Crohn's disease	2	1
Idiopathic	19	9
Positive family history	7	0
Obesity	3	0
Systemic infection and total parenteral nutrition	0	17
Total parenteral nutrition	0	11

- Isebel W, et al. JPGN 2000. 31:411-417

Correlation of age and predisposing factor



- Isebel W, et al. JPGN 2000. 31:411-417

Clinical presentation

- Biliary symptom 52%
- Acute abdominal pain, tenderness with fever 10%
- Nonspecific abdominal pain 24%
- Symptom free 17%

Isebel W, et al. JPGN 2000. 31:411-417

Acute pancreatitis associated with biliary disease in children

Bo H.C, et al. Journal of gastroenterology and hepatology 2003. 18:915-921

Etiology of acute pancreatitis in 56 children

Drug	30%
Biliary disease	29%
Trauma	11%
Systemic disease	9%
Infection	9%
Idiopathic	12%

Bo H.C, et al. Journal of gastroenterology and hepatology 2003. 18:915-921

Acute pancreatitis due to biliary disease in 16 children

- Choledochal cyst 7
- Biliary sludge 6
- Gallstone 2
- Anomalous pancreaticobiliary junction 1

Bo H.C, et al. Journal of gastroenterology and hepatology 2003. 18:915-921

RADIOLOGIC FINDING AND CLINICAL COURSE

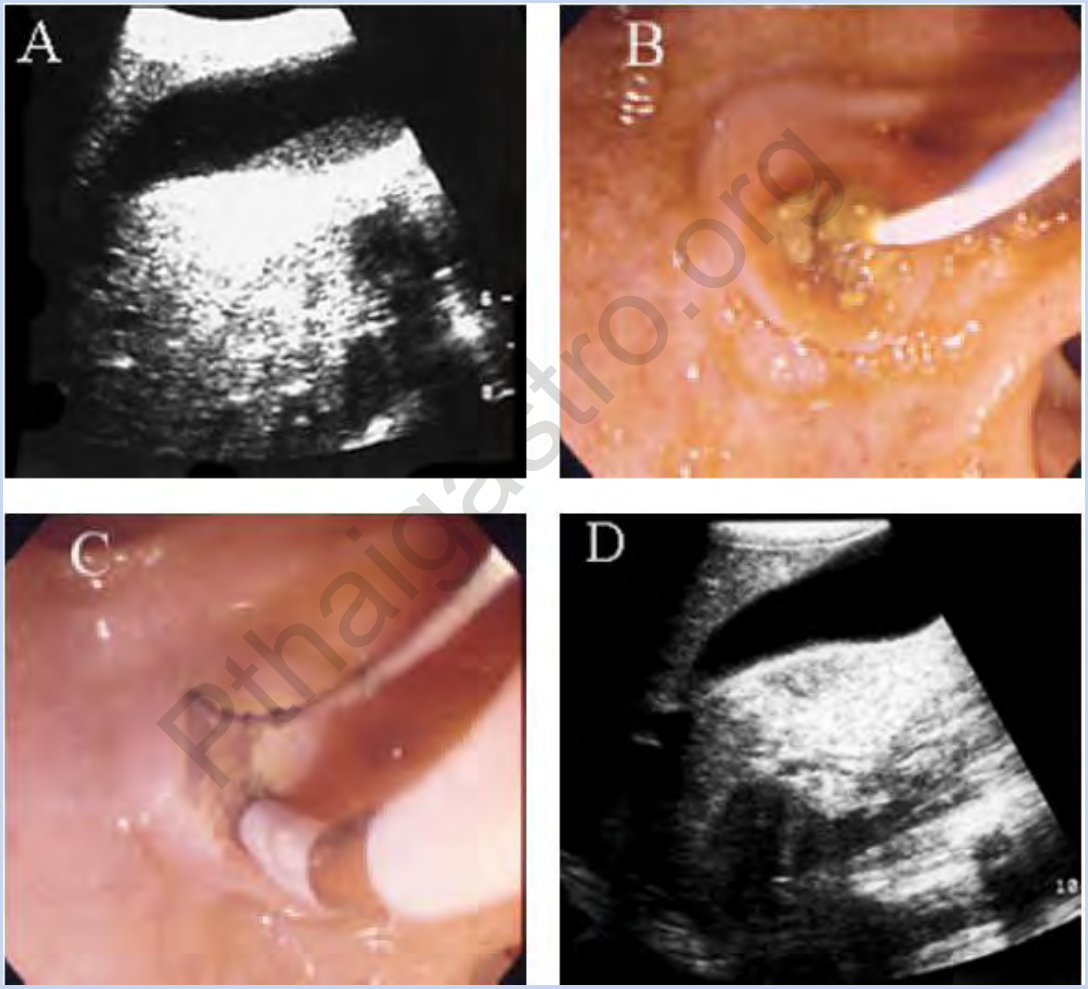
Patient no.	Ultrasound	Computed tomography	ERCP	Operation	Long-term outcome (follow-up period)
1	CDC, normal pancreas	ND	ND	HJS	No recurrence (5 months)
2	CDC, diffuse enlargement of pancreas	ND	ND	HJS	No recurrence (4 months)
3	ND	CDC, diffuse enlargement of pancreas	ND	HJS	No recurrence (4 years)
4	CDC, diffuse enlargement of pancreas, ascites	ND	ND	HJS	No recurrence (4 years)
5	CDC, diffuse enlargement of pancreas	ND	ND	HJS	No recurrence (11 months)
6	CDC, sludge in GB	Enlargement of pancreas head Dilatation of IHD, CBD and PD	APBJ, dilatation of IHD, CBD and PD	HJS	No recurrence (15 months)
7	CDC, sludge in GB, normal pancreas	ND	APBJ, dilatation of CBD, EST	HJS	Recurrence after EST No recurrence after HJS (2 years)
8	Sludge in GB	Normal pancreas	Normal CBD, EST and ENBD	ND	No recurrence (2 months)
9	Sludge in GB and CBD, dilatation of CBD and PD Enlargement of pancreas head	ND	Dilatation of CBD and IHD and PD EST and ENBD	ND	No recurrence (33 months)
10	Sludge in GB and CBD, dilatation of CBD and IHD	Enlargement of pancreas head	Dilatation of CBD and IHD, normal PD EST and ENBD	ND	No recurrence (9 months)
11	Sludge in GB, Dilatation of CBD and IHD	Normal pancreas, dilatation of CBD and IHD	APBJ, dilatation of CBD and IHD EST and ENBD	ND	Recurrence after 2 years No recurrence after 2nd EST (41 months)
12	Sludge in GB	Enlargement of pancreas body and tail	Distended GB EST and ENBD	ND	Recurrence after 3 years Recurrence after 5 years
13	Sludge in GB	Normal pancreas	Dilatation of CBD Distended GB, EST and ENBD	OC	Recurrence after 2 years No recurrence after OC (4 years)
14	Stone in GB	Normal pancreas	ND	ND	Improved spontaneously (1 month)
15	Stone in distal CBD Dilatation of CBD and PD	Normal pancreas	Multiple filling defect in CBD Dilatation of CBD and PD, EST and ENBD	ND	No recurrence after stone extraction (25 months)
16	Normal GB and CBD Diffuse enlargement of pancreas	Diffuse enlargement of pancreas Ascites	APBJ, dilatation of PD Normal GB and CBD	ND	Improved spontaneously (16 months)

Bo H.C, et al. Journal of gastroenterology and hepatology 2003. 18:915-921

RADIOLOGIC FINDING AND CLINICAL COURSE

Sludge in GB	Normal CBD, EST and ENBD	ND	No recurrence (2 month
Sludge in GB and CBD, dilatation of CBD and PD	Dilatation of CBD and IHD and PD EST and ENBD	ND	No recurrence (33 mont
Enlargement of pancreas head			
Sludge in GB and CBD, dilatation of CBD and IHD	Dilatation of CBD and IHD, normal PD EST and ENBD	ND	No recurrence (9 month
Sludge in GB, Dilatation of CBD and IHD	APBJ, dilatation of CBD and IHD EST and ENBD	ND	Recurrence after 2 years No recurrence after 2nd (41 months)
	Distended GB	ND	Recurrence after 3 years
Sludge in GB	EST and ENBD		Recurrence after 5 years
	Dilatation of CBD	OC	Recurrence after 2 years
Sludge in GB	Distended GB, EST and ENBD		No recurrence after OC

Biliary sludge in a child with pancreatitis



Bo H.C, et al. Journal of gastroenterology and hepatology 2003. 18:915-921

BILIARY INDICATION FOR ERCP

Diagnosis	Therapeutic
- Investigation of neonatal cholestasis - Biliary atresia - Choledochol cyst - Choledocholithiasis - Biliary obstruction due to parasitic infestation - Dilated intrahepatic bile duct - Benign and malignant biliary strictures - Primary sclerosing cholangitis - Bile plug syndrome - Manometric evaluation of the sphincter of Oddi	- Sphincterotomy - Sphincteroplasty (balloon dilatation) - Stone extraction - Stricture dilatation - Stent placement - Nasobiliary drainage -Treatment of choledochal cyst -Treatment of traumatic biliary injuries

Lower M, et al. Walker's Pediatric Gastrointestinal Disease 5th ed, 2008

Acute pancreatitis

- Definition, diagnosis
- Assessment severity : CT severity index, Ranson criteria, APACHE II score
- Treatment major symptom (pain, nausea and vomiting, hypovolemia)
- Limiting progression

Severe pancreatitis

- One of the following
- Presence of local complications

  pancreatic necrosis, pseudocyst, abscess
- Presence of organ failure

  shock, pulmonary insufficiency, renal insufficiency, GI bleeding > 500 ml in 24 hr
- >/= 3 Ranson's criteria
- >/= 8 APACHE II point

CT SEVERITY INDEX (CTSI)

CT grade score		Necrosis score	
A	0	None	0
B	1	<33%	2
B	2	35-50%	4
D	3	>50%	6
E	4		

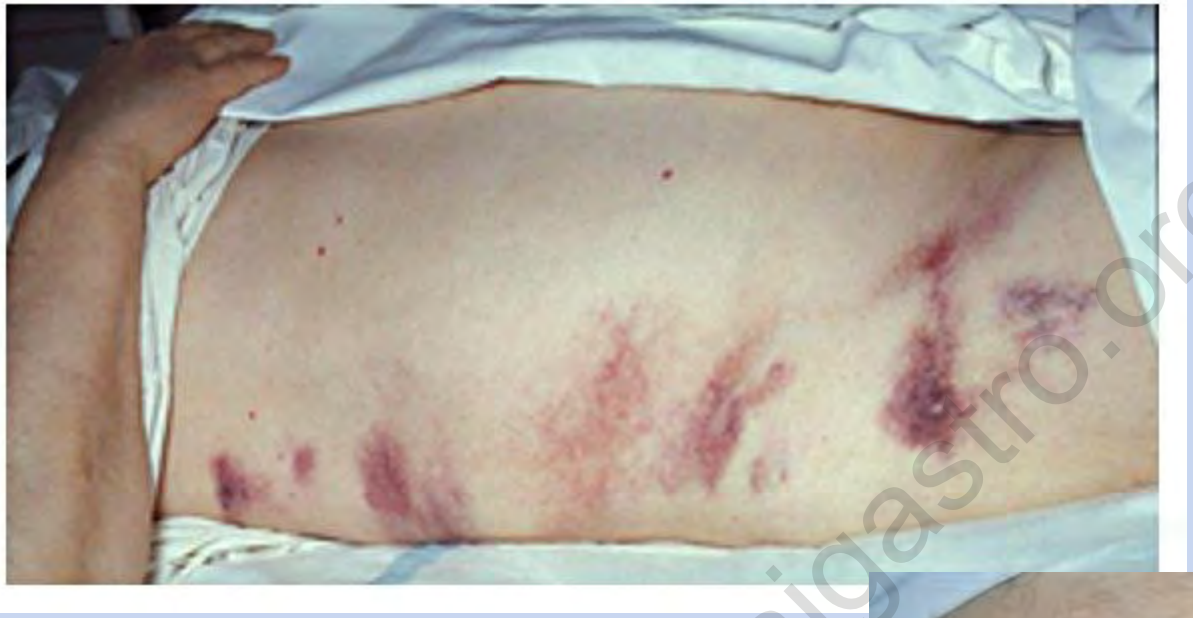
- Grade A : normal pancreas
- Grade B : pancreatic enlargement
- Grade C : inflammation pancreas, panceatic fat
- Grade D : single peripancreatitis fluid collection
- Grade E : two or more fluid collection or retroperitoneal air

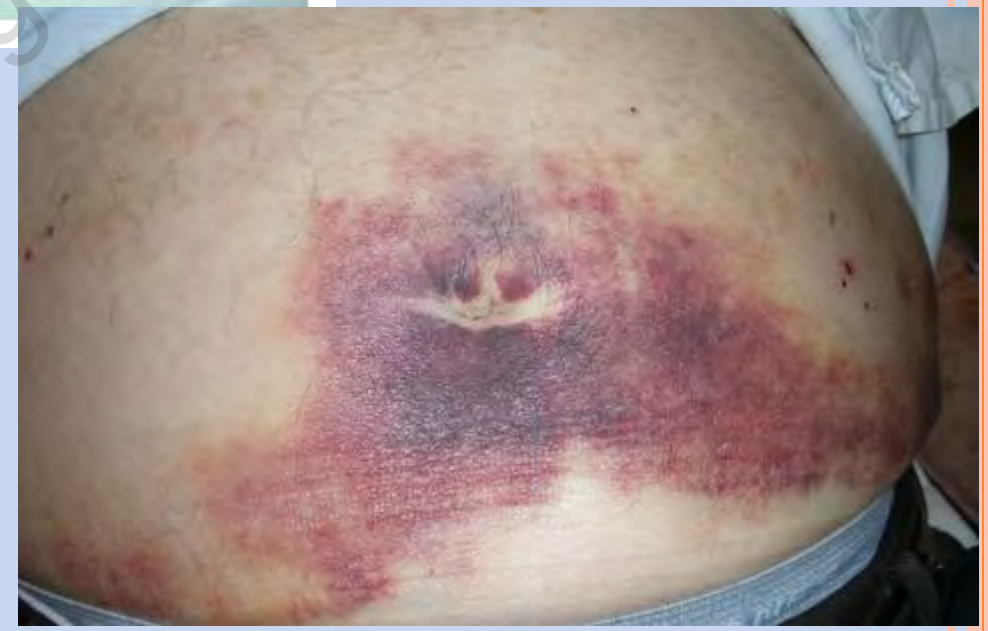
Ranson's criteria for acute gallstone pancreatitis

At admission		During the initial 48 hr	
Age	> 70 year	Hct fall	> 10 %
WBC	> 18,000	BUN elevation	> 2 mg/dl
Blood sugar	> 220 mg/dl	Serum calcium	< 8 mg/l
Serum LDH	> 400 IU/L	Base deficit	> 5 mEg/l
Serum AST	> 250 U/dl	Estimate fluid sequestration	> 4 L

- Ranson score — mortality rate
- 0-2 — 2%
- 3-4 — 15%
- 5-6 — 40%
- 7-8 — 100%

Turner's sign, Cullen's sign





Management

- Pain
- Fluid and electrolyte supplement
- Role of nasogastric decompression
- Prophylactic antibiotics
- Nutrition support

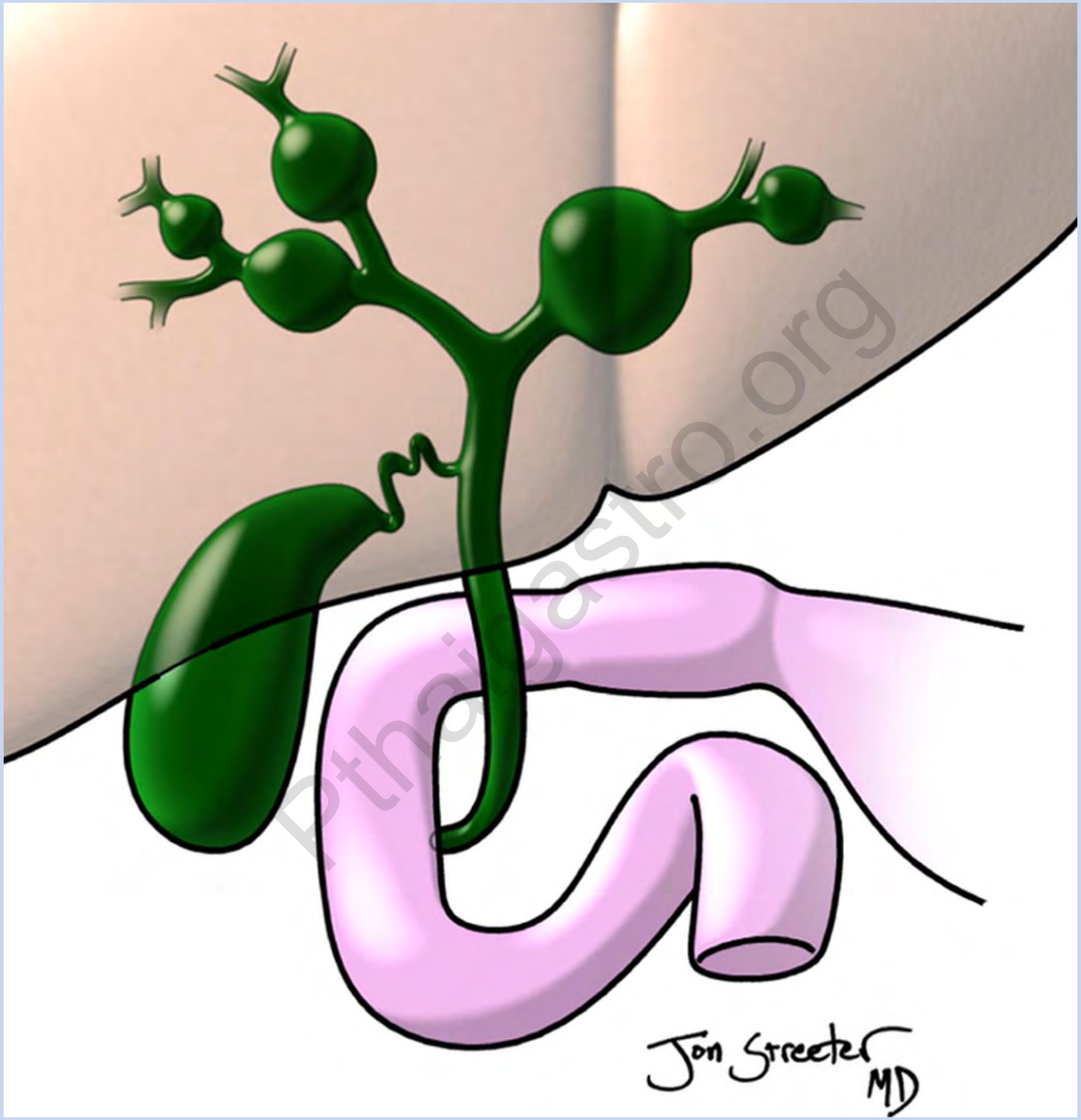
Hyperlipoproteinemia

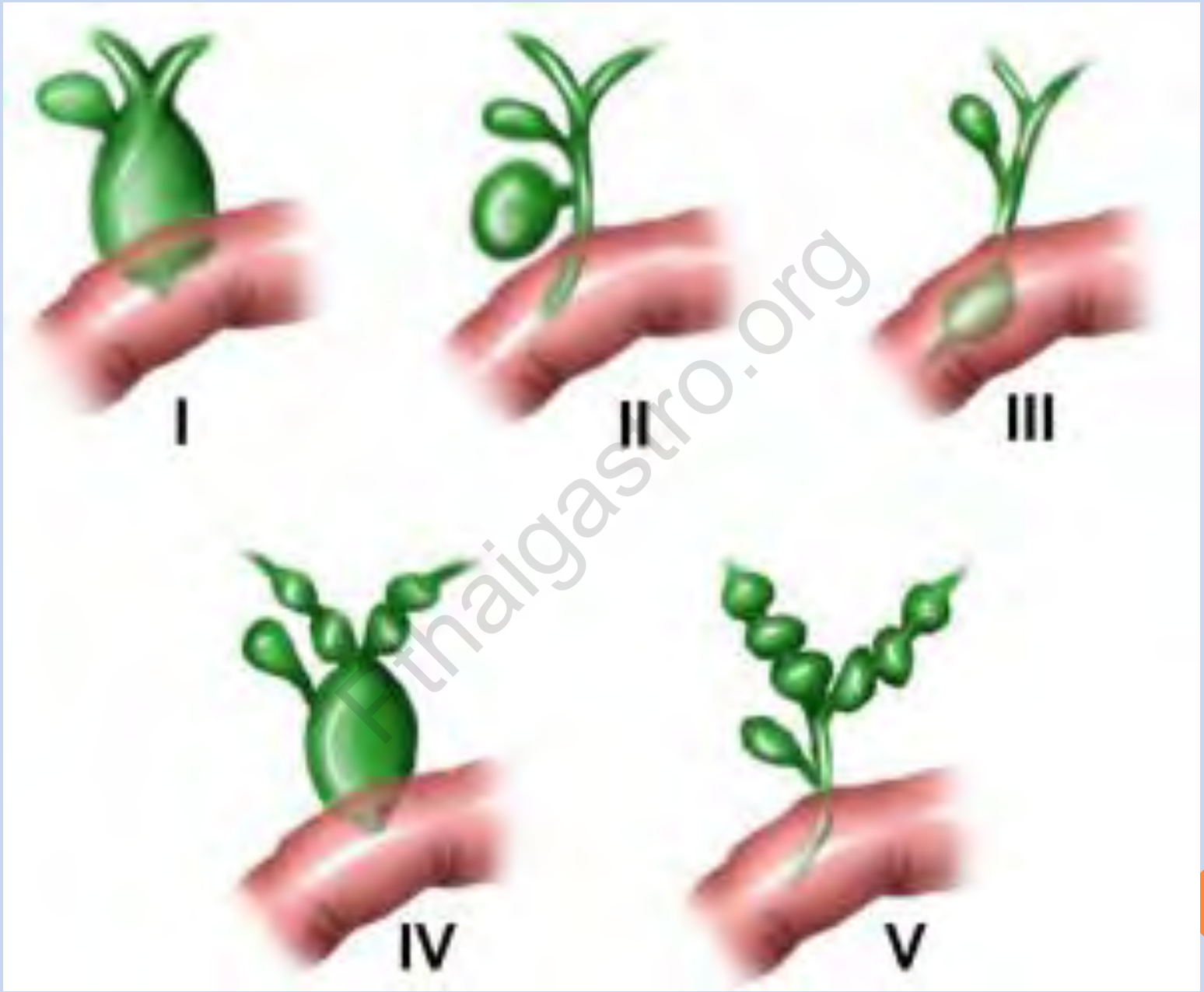
- Primary causes : gene defects
- Secondary causes

  - glycogen storage disease
  - congenital biliary atresia
  - hypothyroidism
  - nephrotic syndrome
  - diabetes mellitus
  - obesity
  - drugs : prednisolone, anabolic steroids, oral contraceptives

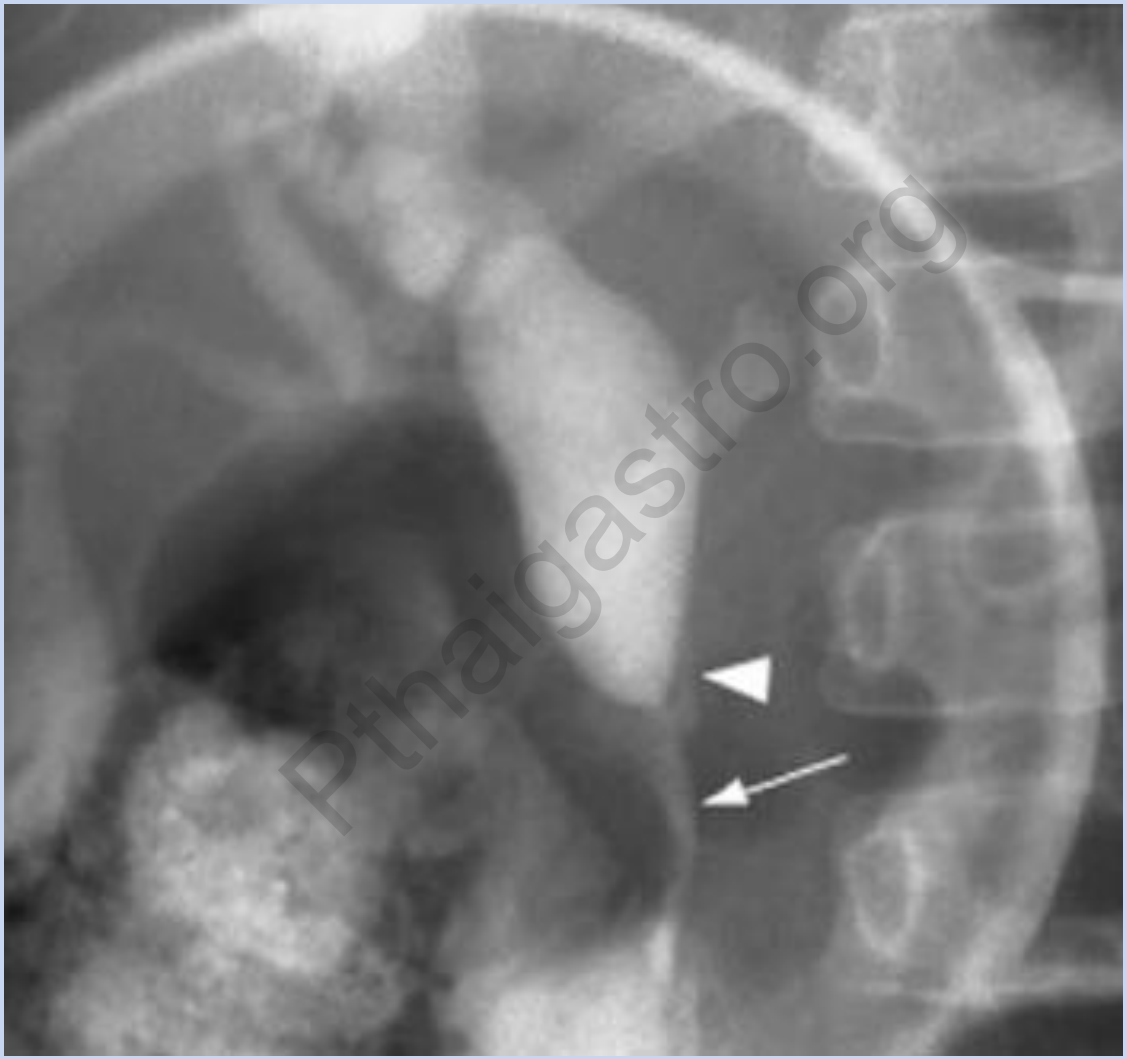
FAMILIAL HYPERLIPOPROTEINEMIA

type	Elevated particle	Major abn. Lipid	frequency	cause
I	Chylomicron	Very high TG	Very rare	LPL , apoC- II gene mutation
IIA	LDL	High Chol.	Common	LDLR, apoB100 gene mutation
IIB	LDL, VLDL	High chol. + TG	Common	Unknown
III	IDL	High chol. + TG	Rare	ApoE2 polymorphism + additional factors
IV	VLDL	High TG	Common	apoC-III gene mutation
V	Chylomicrons, VLDV	Very high TG	rare	Lipid overload, insulin resistance, apoC-100 gene mutation





ERCP showing type IV choledochal cyst



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