



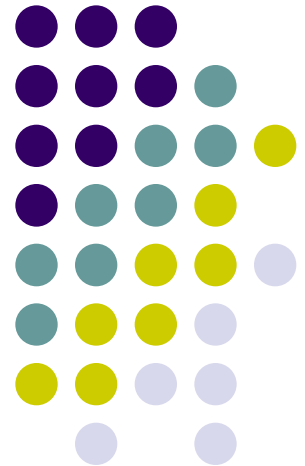
Pediatric GI Interhospital Conference

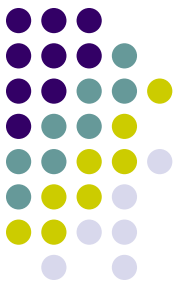
พญ.ศิริลักษณ์ เจนนุวัตร

พญ.นิยะดา วิทยาศัย

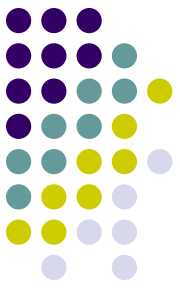
งานโรคระบบทางเดินอาหารและโภชนาการคลินิก

กลุ่มงานกุมารเวชศาสตร์ สถาบันสุขภาพเด็กแห่งชาติมหาราชินี





- เด็กชายไทย อายุ 3 ปี ภูมิลำเนา กรุงเทพฯ
- ประวัติจากมารดา
- CC: ปวดท้อง 3 สัปดาห์
- PI: 3 สัปดาห์ ก่อนมาร.พ. มีอาการปวดท้อง บอกตำแหน่งที่ปวดไม่ได้ เวลาปวดจะร้องว่าปวดท้อง แล้วนอนตัวงอ เป็นประมาณ 2-3 ชั่วโมง มีคลื่นไส้อาเจียน เป็นอาหารที่ทานเข้าไป ไม่มีสีเขียว ไม่มีเลือดปน 1 ครั้ง /วัน เวลาหายปวดจะปกติ กินได้ วิ่งเล่นได้



- ไม่มีไข้ ไม่มีไอ ไม่มีน้ำมูกมาก่อน ไม่เคยมีตัวเหลืองตาเหลือง ถ่ายทุกวัน วันละครึ่ง ไม่มีถ่ายเหลว ไม่มีมูกเลือด ลักษณะถ่ายเป็นก้อนสีเหลือง ปัสสาวะเหลืองใส ไม่แสบขัด
- มาสถาบันสุขภาพเด็กฯ ตรวจร่างกาย abdomen: soft, not tender ได้ air-x drop กลับบ้านไป ไม่ปวดท้อง กินได้ เล่นดี

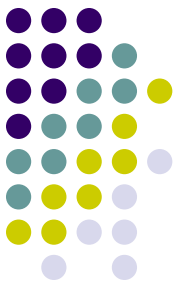


- 1 สัปดาห์ก่อน มีอาการปวดท้องแบบเดิม ไม่มีไข้ มา สถาบันสุขภาพเด็กฯ Dx: viral gastritis, Rx: motilium ORS กลับบ้าน
- หลังกลับบ้านไป ยังมีอาการปวดท้องอีก น้ำหนักลด 1 kg. ใน 3 สัปดาห์จึงมา สถาบันสุขภาพเด็กฯ อีกครั้ง ตรวจร่างกาย mild tender at epigastrium, hyperactive bowel sound
- UA: wbc 0-1, rbc-neg
- Rx: air-x , นัด F/U GI clinic



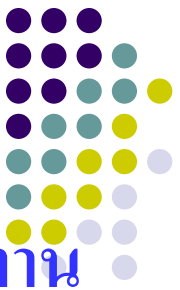
- 3 วันก่อน ปวดท้องมากขึ้น ต้องอุ้มตลอด มีอาเจียนเป็นอาหารที่กินเข้าไป ไปนอน รพ.เอกชน อาการไม่ดีขึ้น จึงมา สถาบันสุขภาพเด็กฯ

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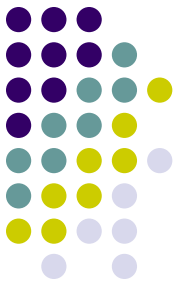
Past history

- บุตร 2/2 คลอดครบกำหนด C/S due to previous C/S, BW 3,410 g., ไม่มีปัญหาหลังคลอด
- ปฏิเสธโรคประจำตัว ไม่เคยแพ้ยา ไม่เคยแพ้อาหาร
- 1 ปีก่อน เคยมีอาการปวดท้องเป็นๆ หายๆ เป็น 3 ครั้ง, ล่าสุดเป็นเมื่อประมาณ 4 เดือนก่อน เป็นอยู่ 2 วัน ไป clinic ได้ยาแก้ปวด ท้องมากขึ้นอาการดีขึ้น ไม่เคยนอน ร.พ.

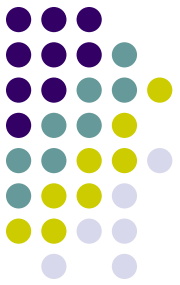


- **Nutrition:** กินข้าว 3 มื้อ แต่ไม่ตรงเวลา ทานเนื้อสัตว์ ชอบทานผักผลไม้ กินขนมถุง ถุงละ 6 บาท 1-2 ถุง/วัน นมเปรี้ยว /ยาคูลท์ 1-2 ขวด/วัน
- **Vaccine:** ครบ
- **G&D:** ขึ้นบันไดสลับขา, คอยเก่ง เล่าเรื่องตัวเองให้ฟังได้เข้าใจ, เล่นกับคนอื่นได้
- **FH:** ปฏิเสธโรคกระเพาะ, โรคทางเดินอาหารในครอบครัว
- **บิดา มารดา** ไม่มีโรคประจำตัว
- **ENV:** อยู่บ้าน 4 คน เลี้ยงปลา ไม่มีใครในบ้านสูบบุหรี่

Physical examination

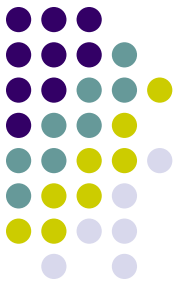


- **V/S : T 37°C, RR 32/min, PR 104 bpm, BP 118/72 mmHg**
- **BW 13 kg., Ht 98 cm.**
- **W/A 89.6%, W/H 86.6%, H/A 103%**
- **Good consciousness, not pale, no jaundice, no dry lip, no sunken eyeball**
- **Pharynx and tonsils: not injected, no dental careis**



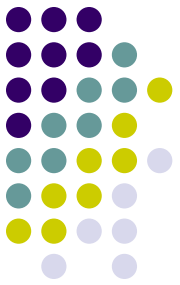
- **Heart: normal s_1, s_2 , no murmur**
- **Lungs: clear**
- **Abdomen: soft, mild distension, not tender, impalpable liver and spleen**
- **Neuro: WNL**

Problem list



- **Chronic abdominal pain**
- **Acute mild PEM**
- **Weight loss**

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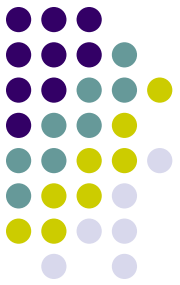
Investigation

- **Hb 12.0 g/dl, Hct 38.9 % WBC 14,600 /ul (N 46%, L 47%, M 4%, E 3%) , Plt 423,000**
- **Electrolyte: Na 135 mmol/L, K 3.96 mmol/L, Cl 104 mmol/L, CO₂ 20.3 mmol/L**

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Differential diagnosis

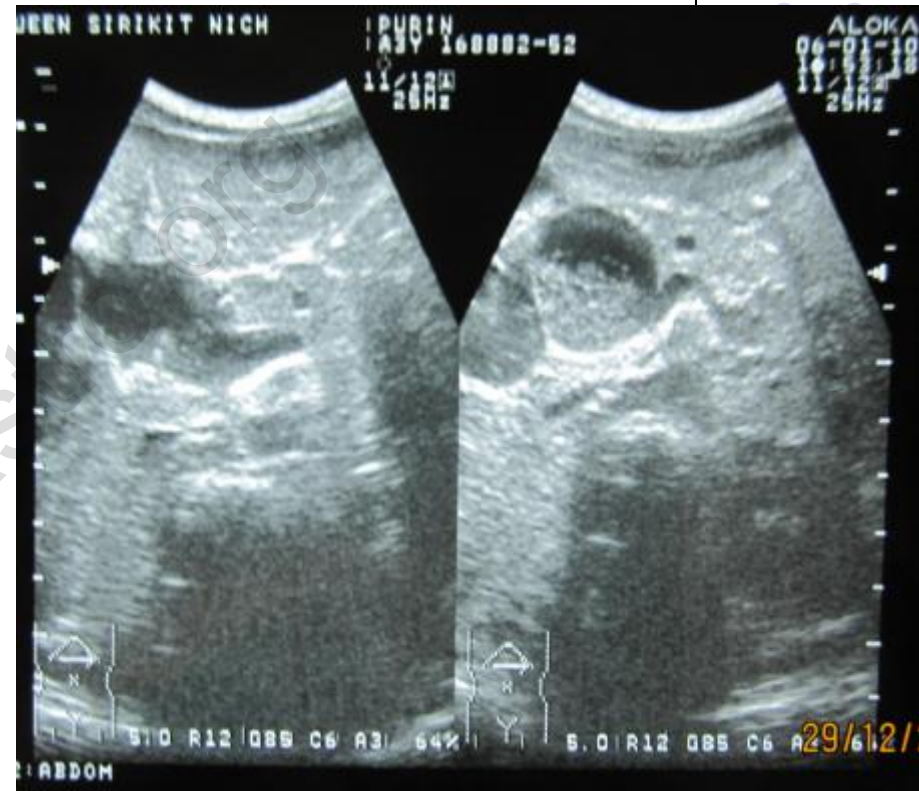


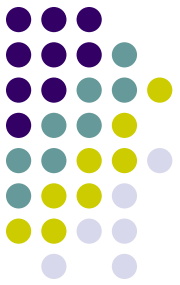
- **Medical condition**
 - Peptic ulcer
 - Pancreatitis
 - Hepatitis
- **Surgical condition**
 - Intussusception
 - Choledochal cyst

Pathologygastro.org





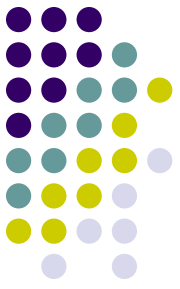




U/S upper abdomen

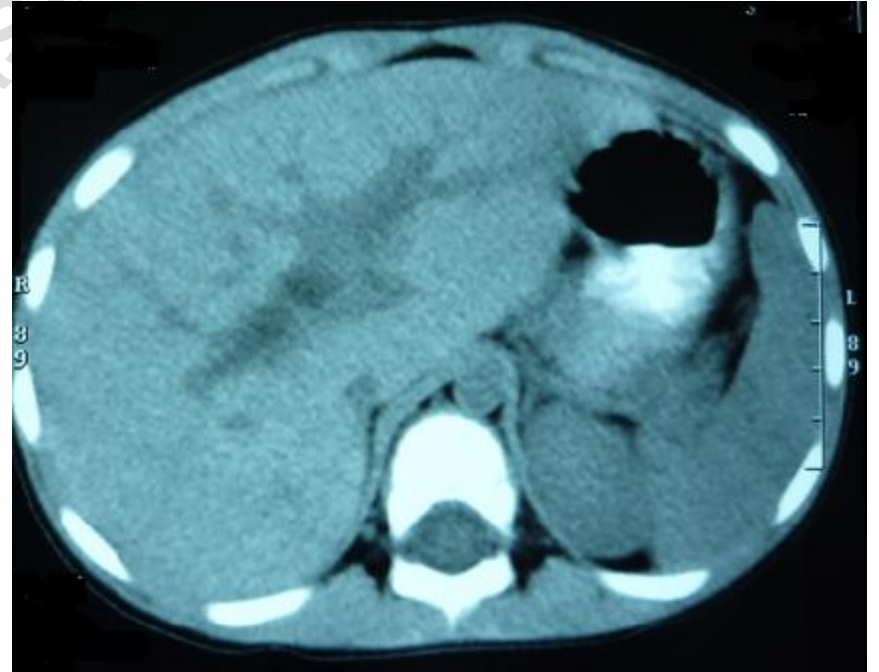
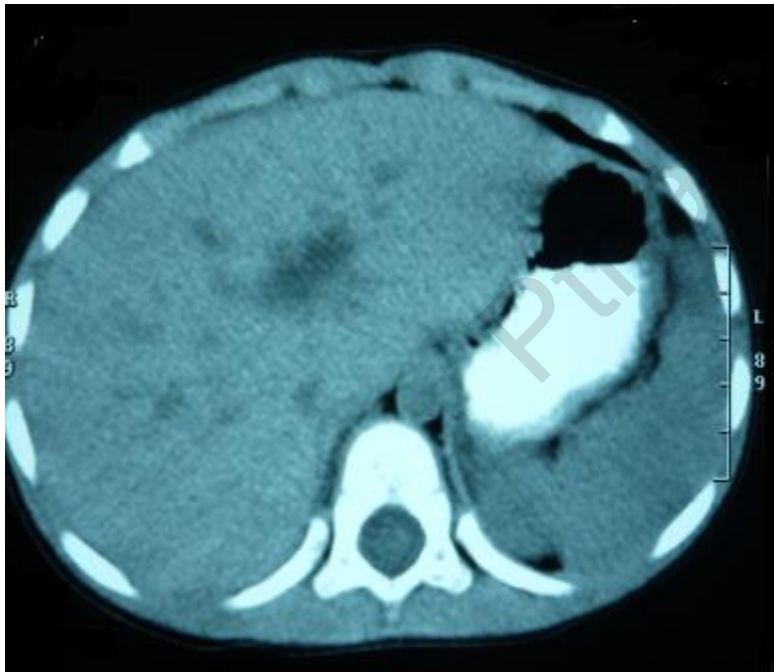
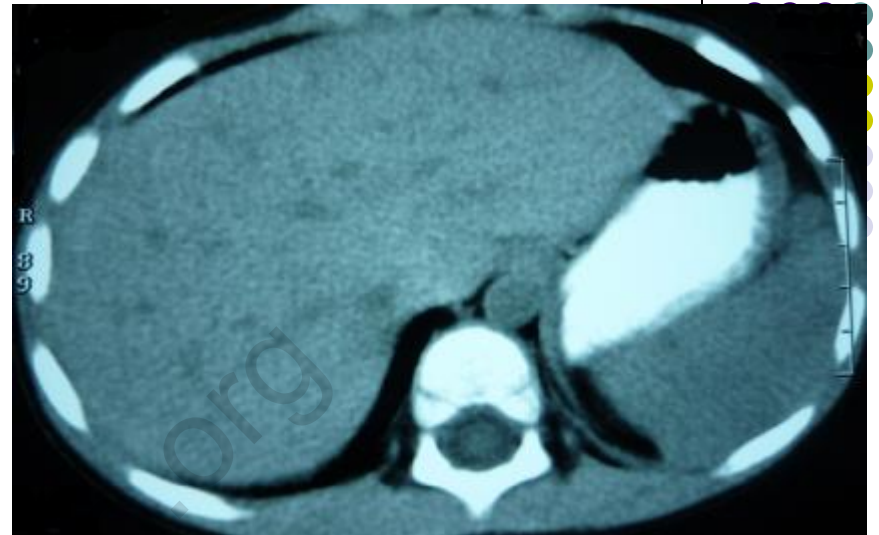
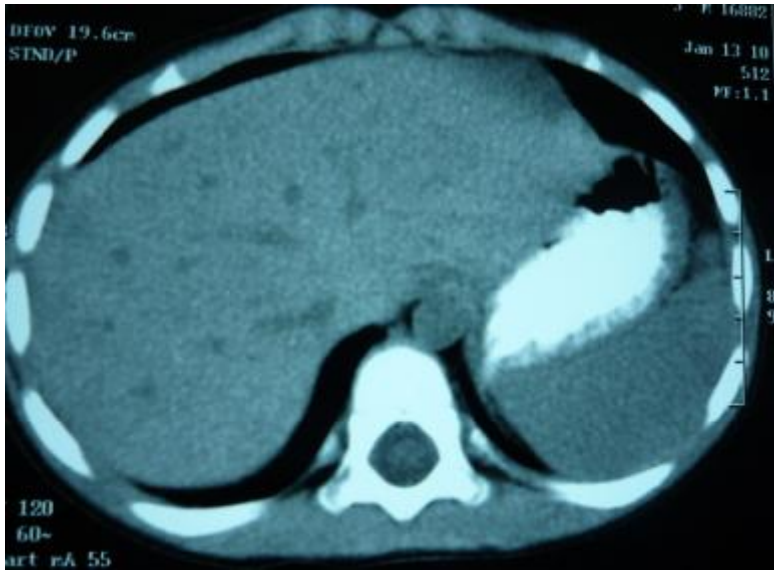
IMP : Choledochal cyst with intrahepatic duct dilatation. Turbid bile is seen. Mild hepatomegaly and parenchymal disease is not excluded.

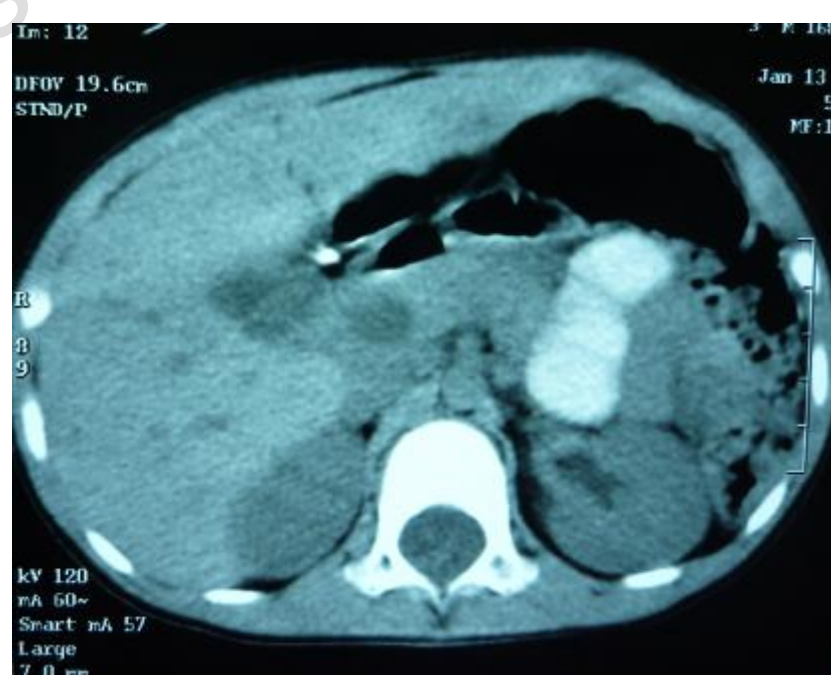
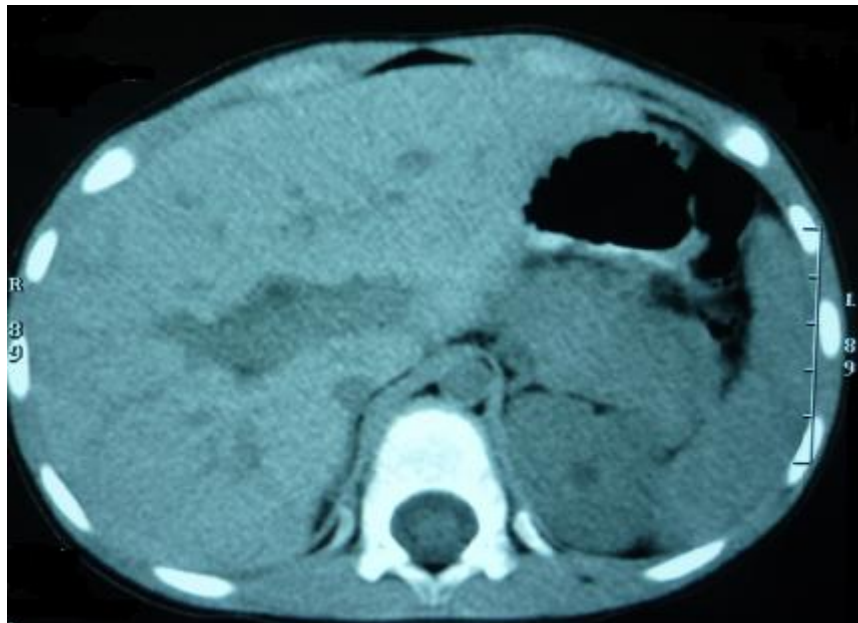
Prominent pancreatic size and slight increased parenchymal echogenicity. Pancreatitis is not excluded

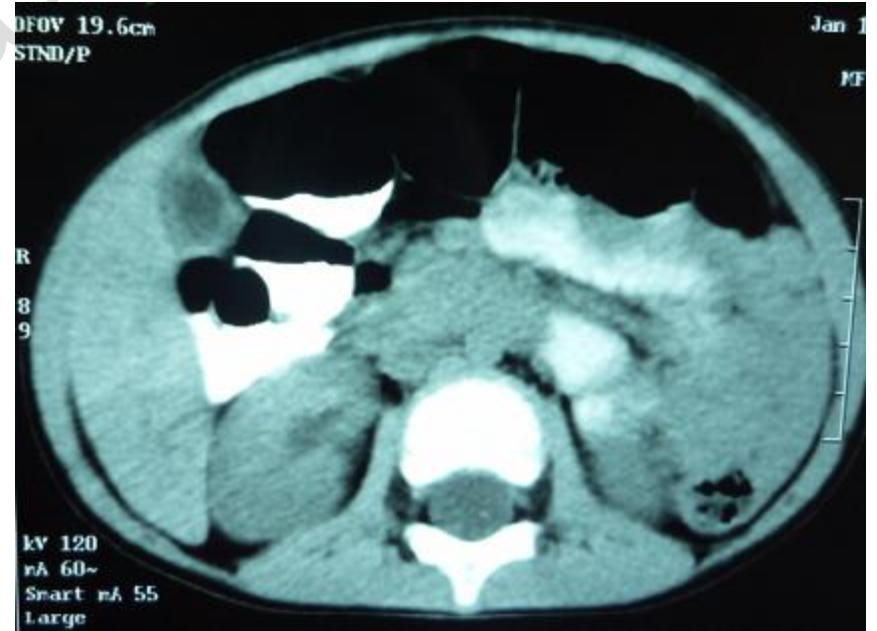
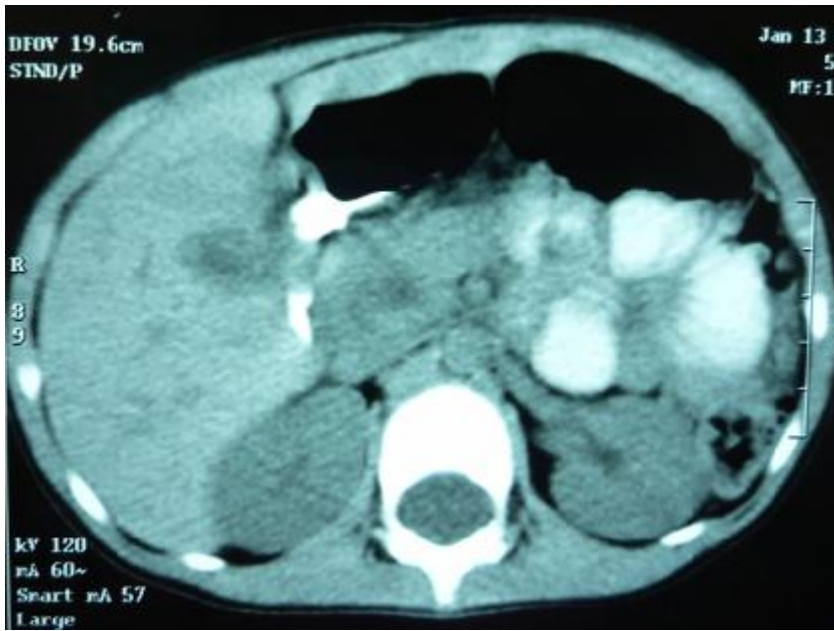


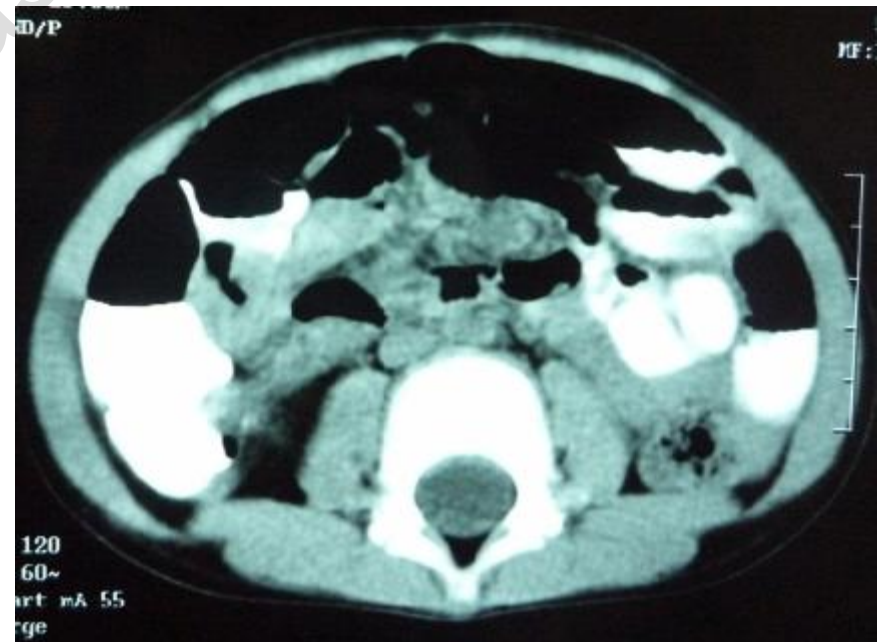
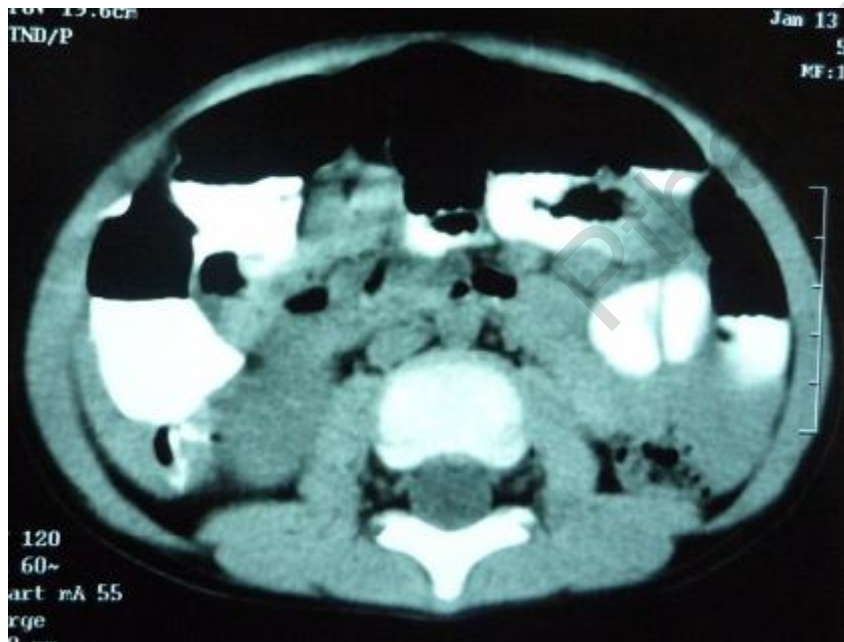
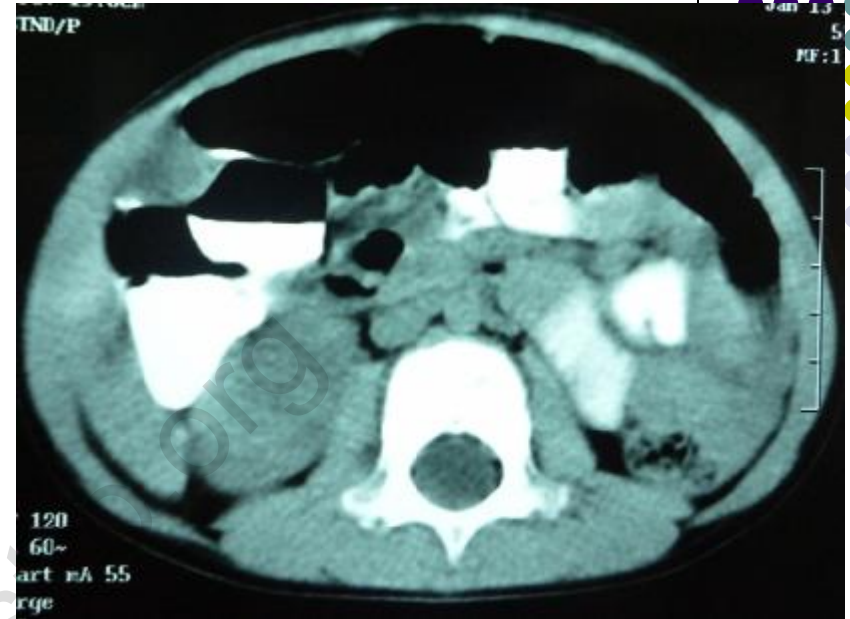
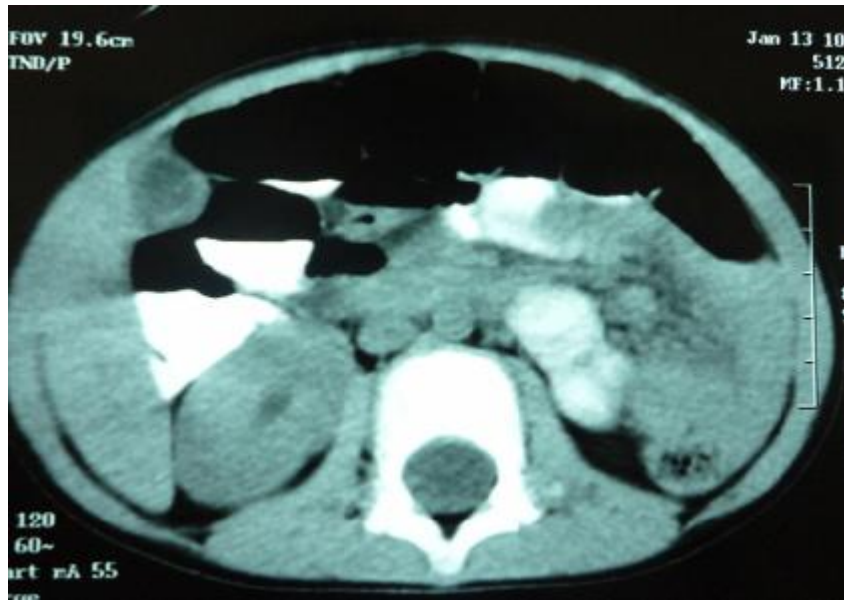
Investigation

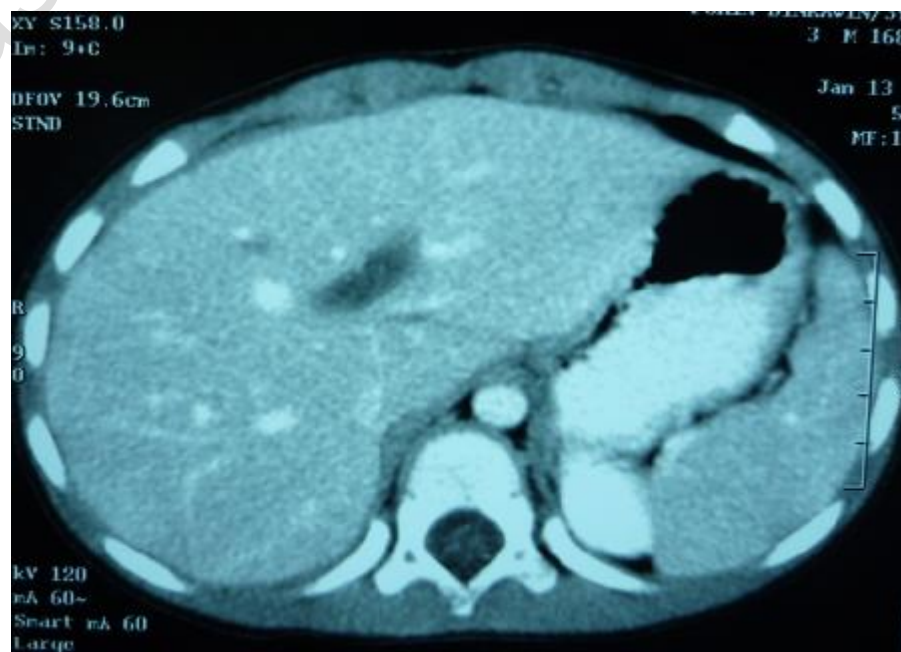
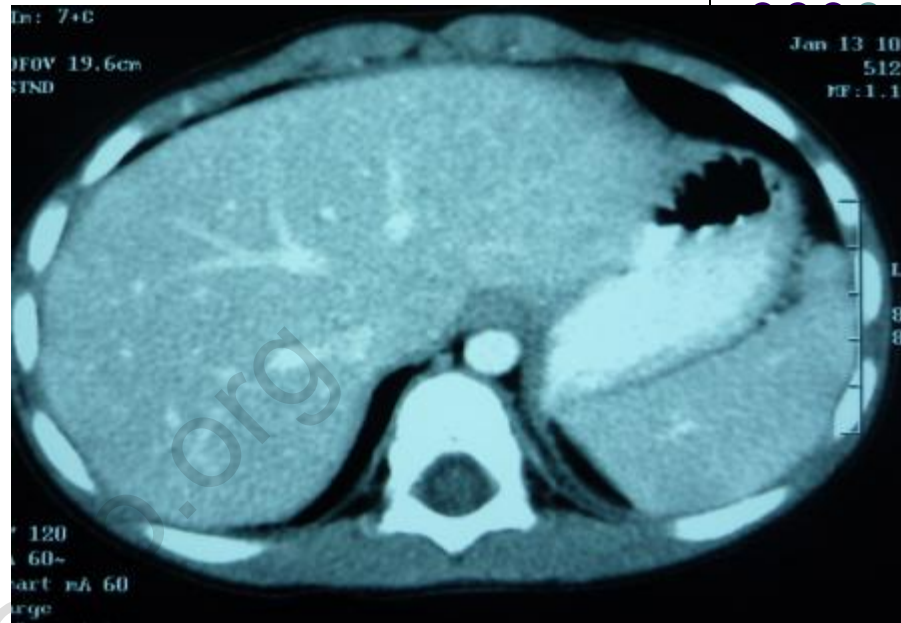
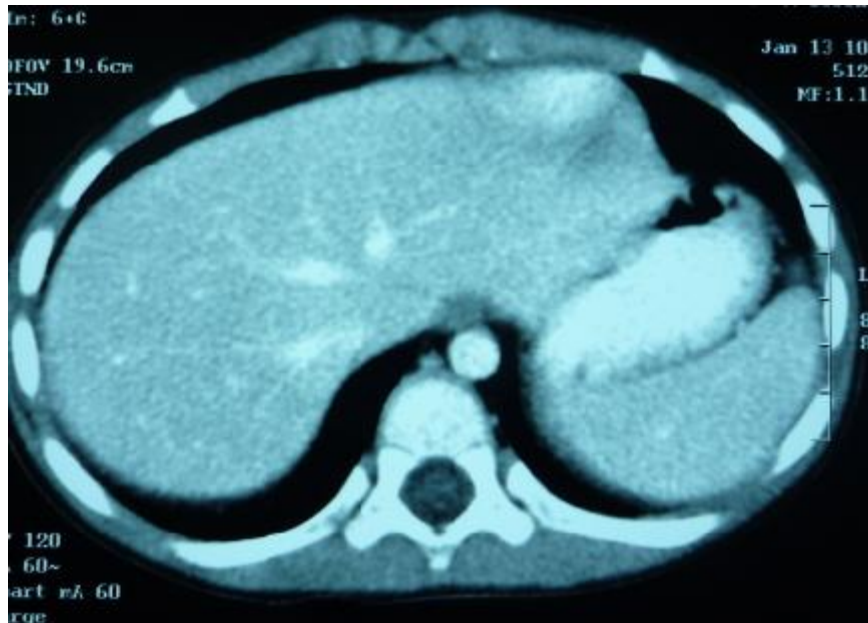
- **LFT : TP 6.51, Alb 4.15, Glob 2.36 g/dl,
Chol 169 mg/dl, TB 3.58, DB 1.46, IB 2.12 mg/dl,
AST 245, ALT 131, ALP 330 U/L**
- **Amylase 909 U/L**

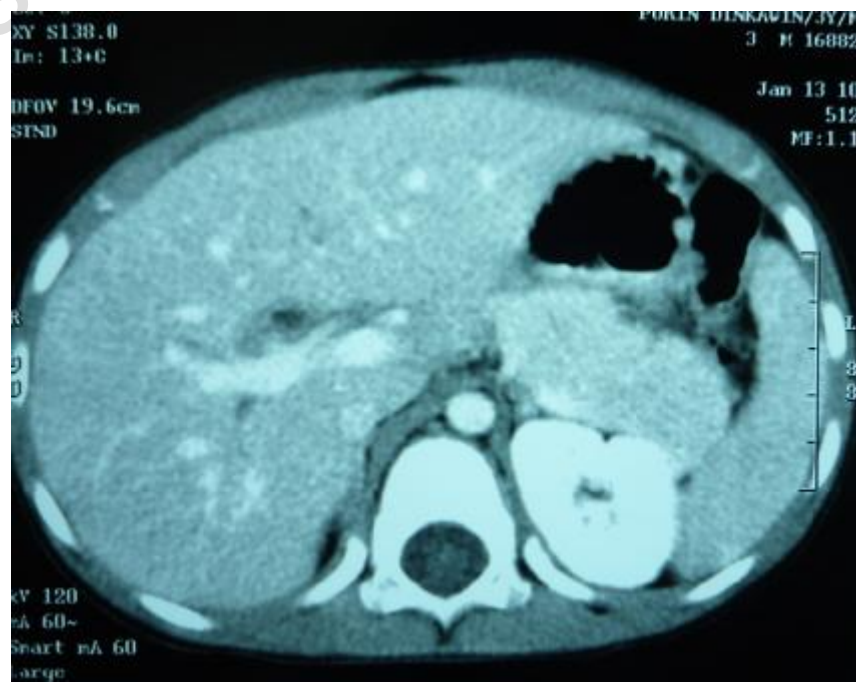
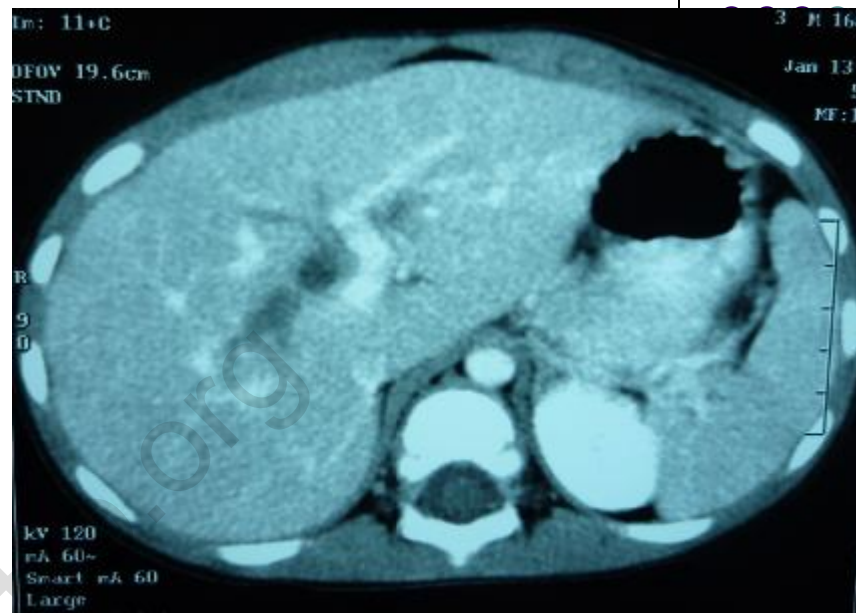
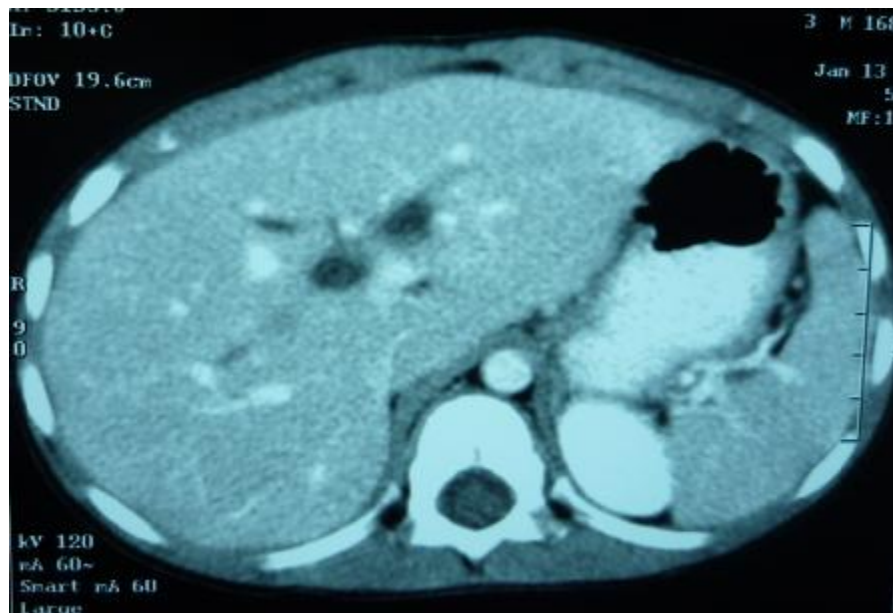


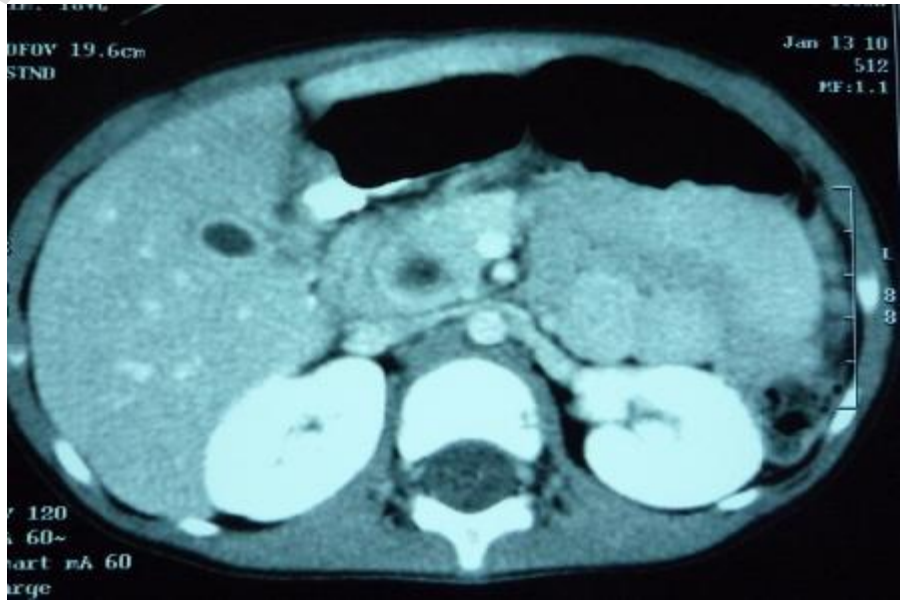
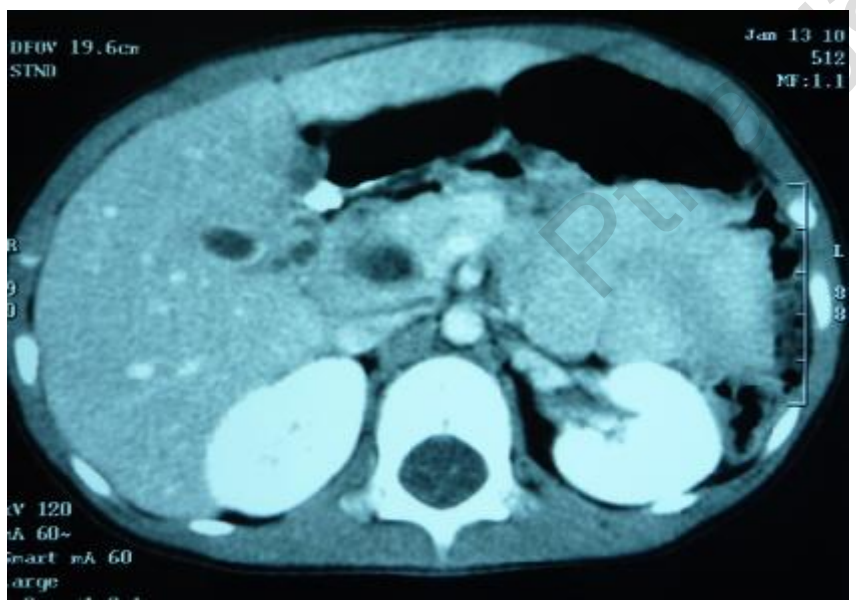
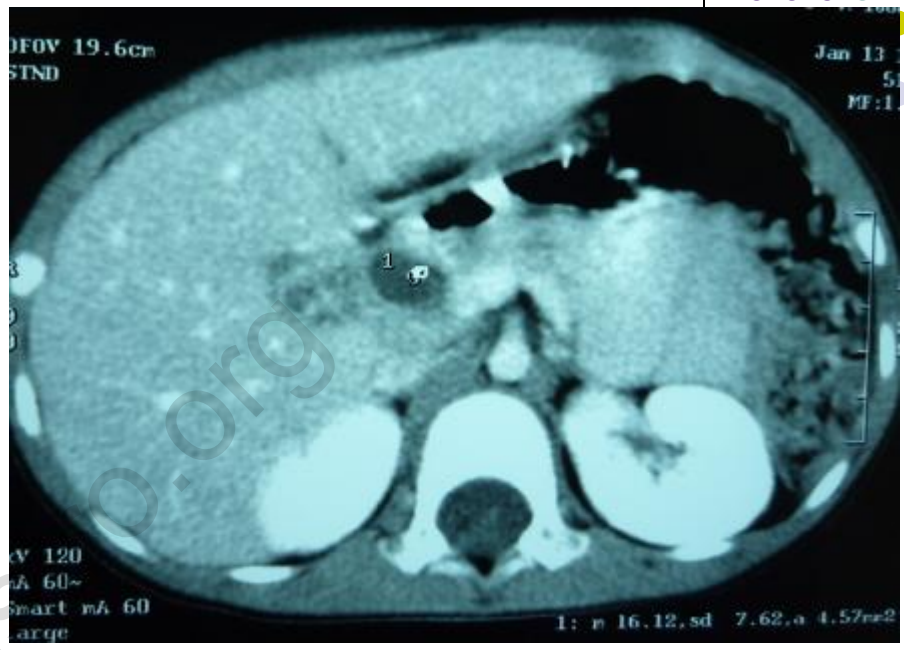


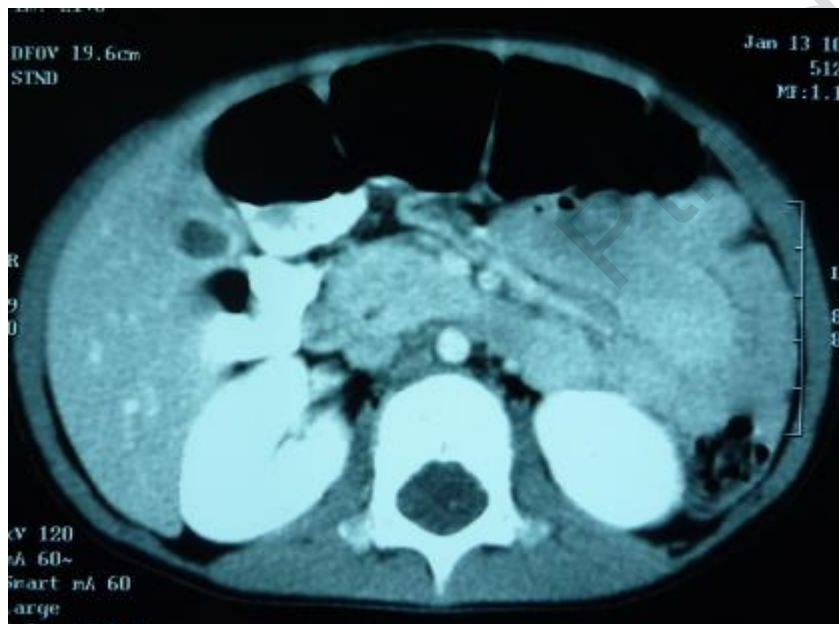
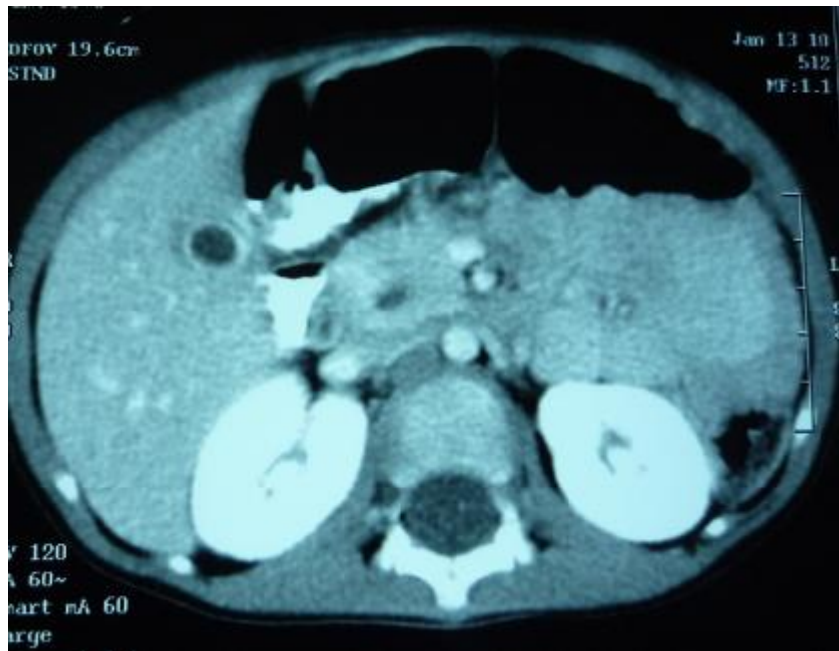


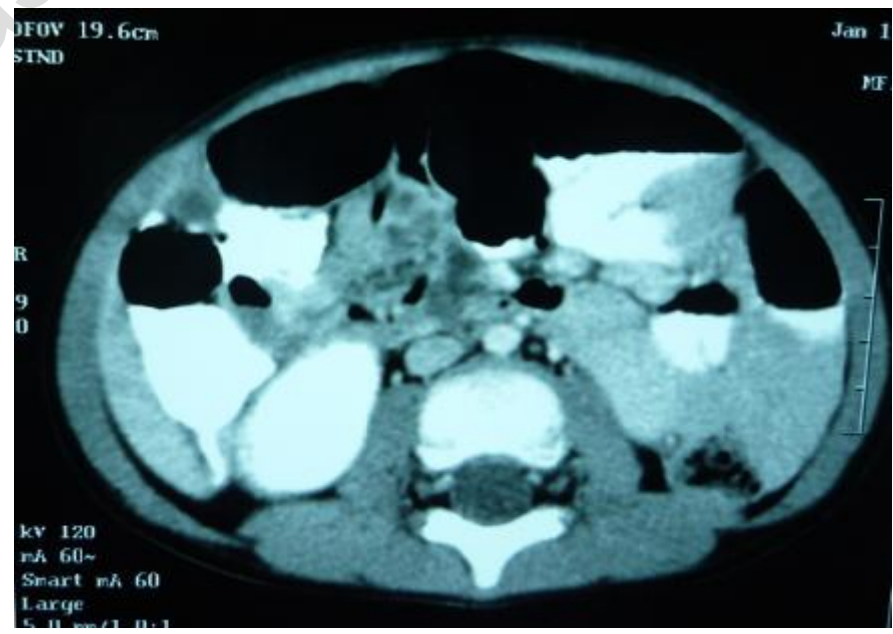
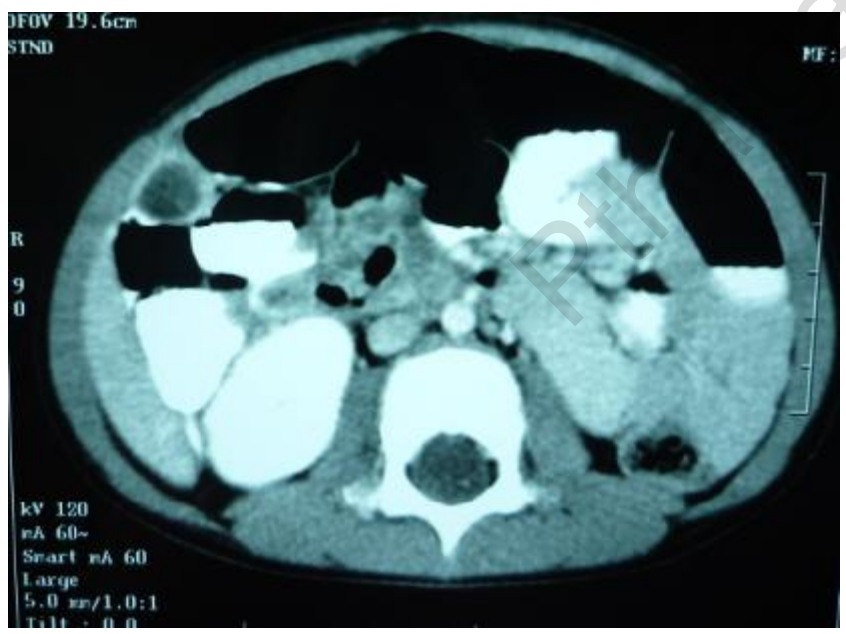
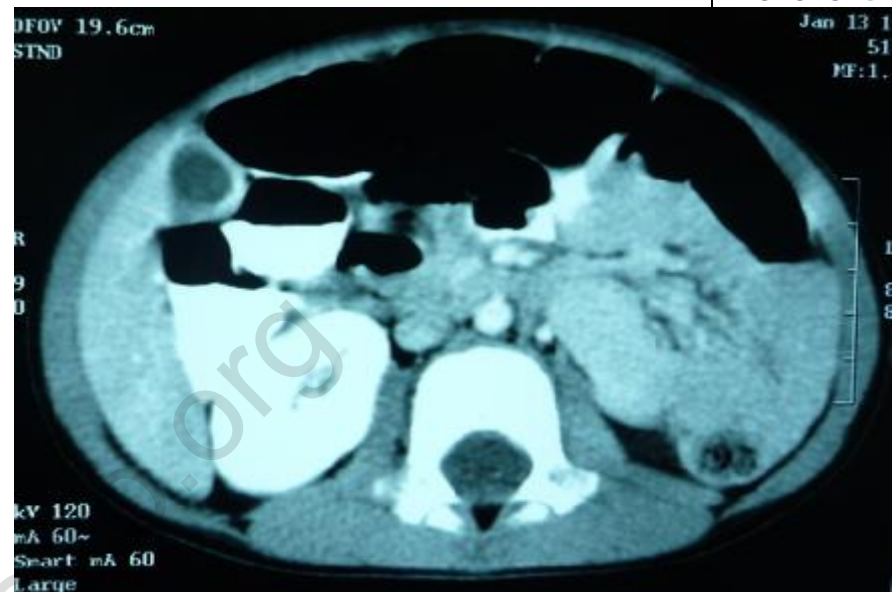
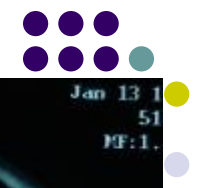


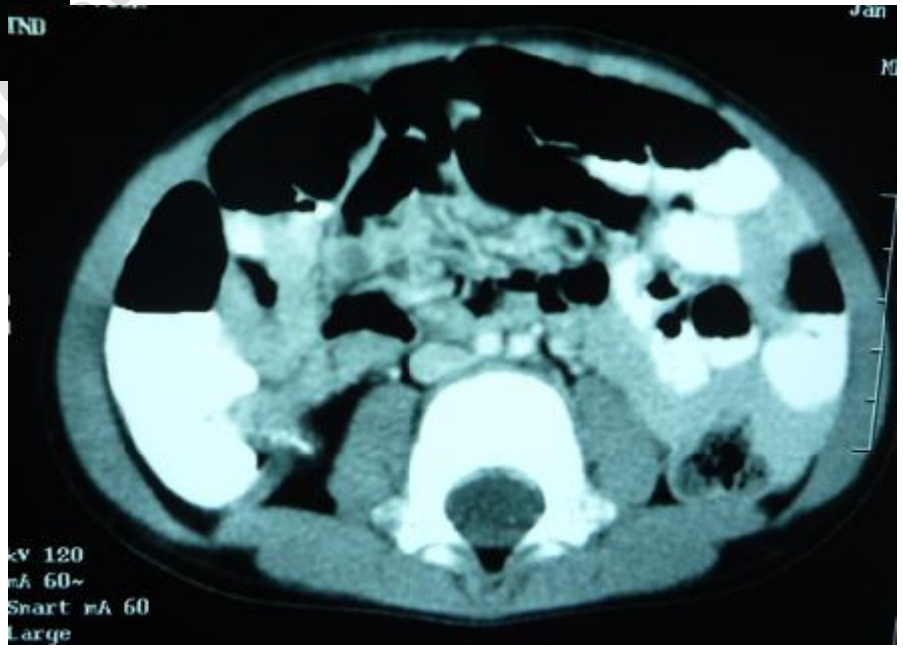






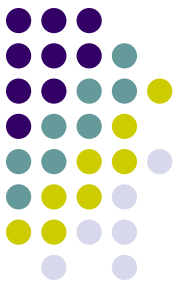






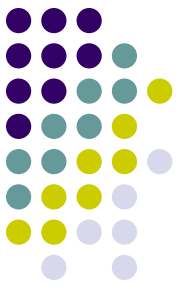
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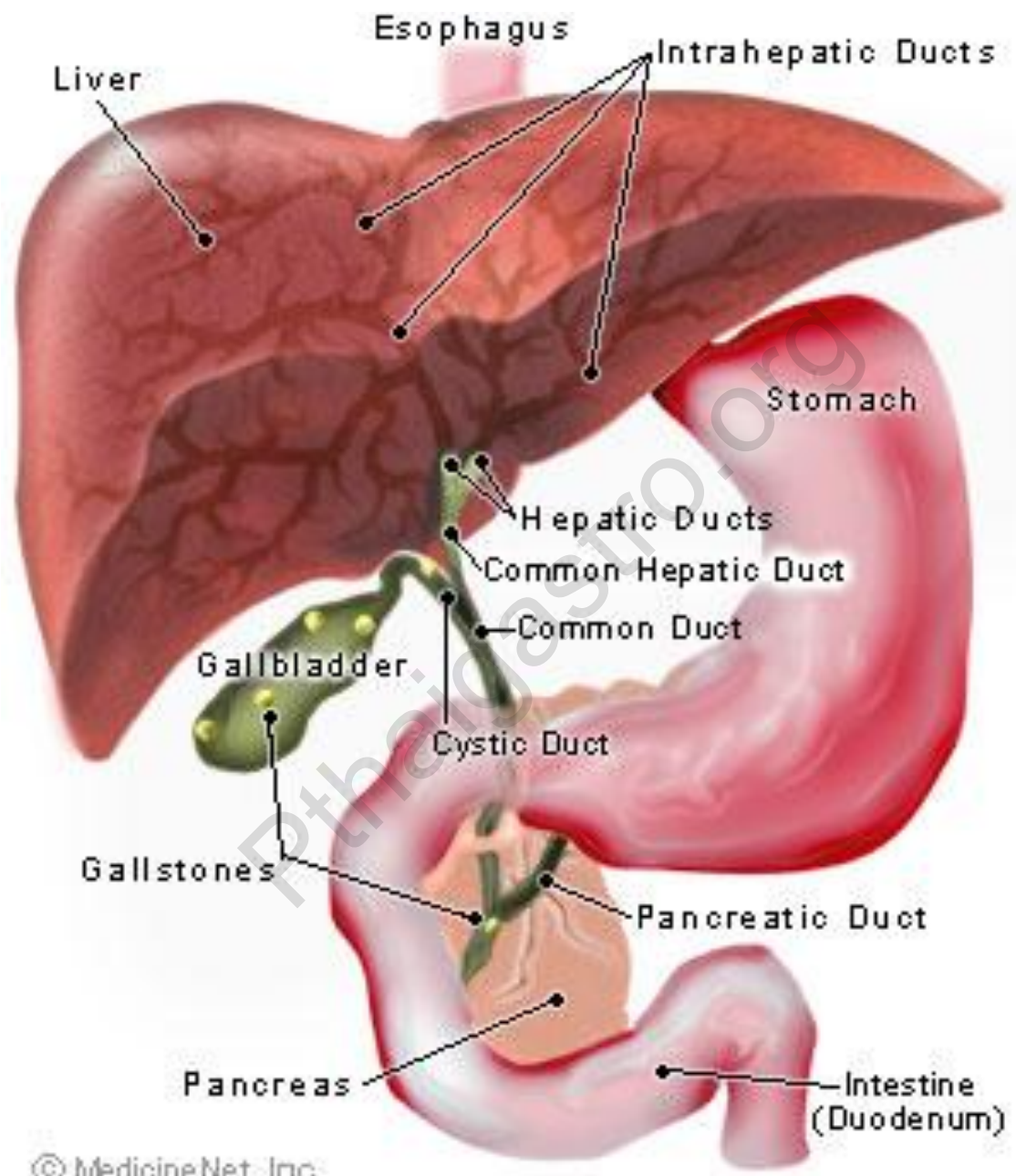
CT Abdomen

- **Dilatation of CBD and intrahepatic ducts are seen down to pancreatic head.**
- **No SOL in liver is seen**
- **GB shows prominent and mild enhancing wall, DDX cholecystitis**
- **Pancreas shows prominent size and mild proximal intrapancreatic duct dilatation. Mild irregular outline and slightly increased density of tissue around the pancreas are seen**
No mass, calcification or cyst formation is seen.
- **Spleen and both kidneys are unremarkable.**
- **No free fluid is seen.**



Choledochal cyst

- **Congenital anomalies of biliary tract characterized by varying degrees of cystic dilatation at various segments of biliary tract (extrahepatic or intrahepatic)**
- **1:15,000 live births in Western**
- **1:1,000 live births in Japan**
- **Female:male 4:1**



Feldman: Sleisenger & Fordtran's Gastrointestinal and Liver Disease, 8th ed.

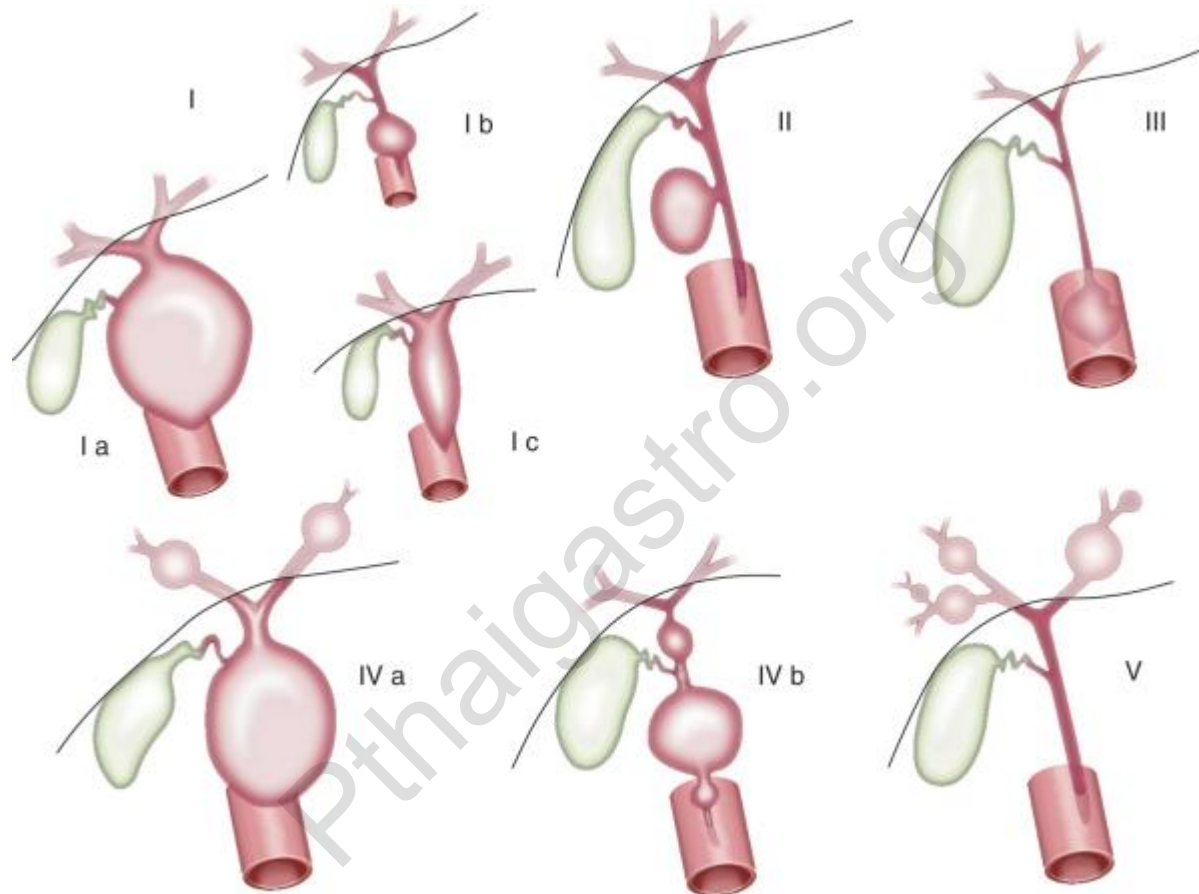
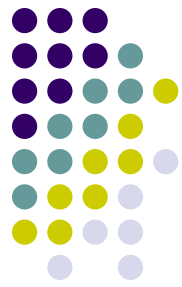
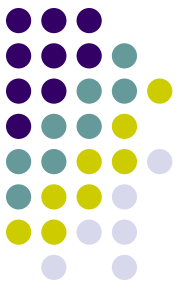
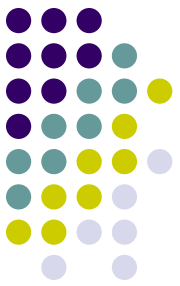


Figure 59-6 Classification of choledochal cysts according to Todani and colleagues.^[105] Ia, common type; Ib, segmental dilatation; Ic, diffuse dilatation; II, diverticulum; III, choledochocele; IVa, multiple cysts (intrahepatic and extrahepatic); IVb, multiple cysts (extrahepatic); V, single or multiple dilations of the intrahepatic ducts. (From Savader SJ, Benenati JF, Venbrux AC, et al: *Choledochal cysts: Classification and cholangiographic appearance. AJR Am J Roentgenol* 156:327, 1991.)



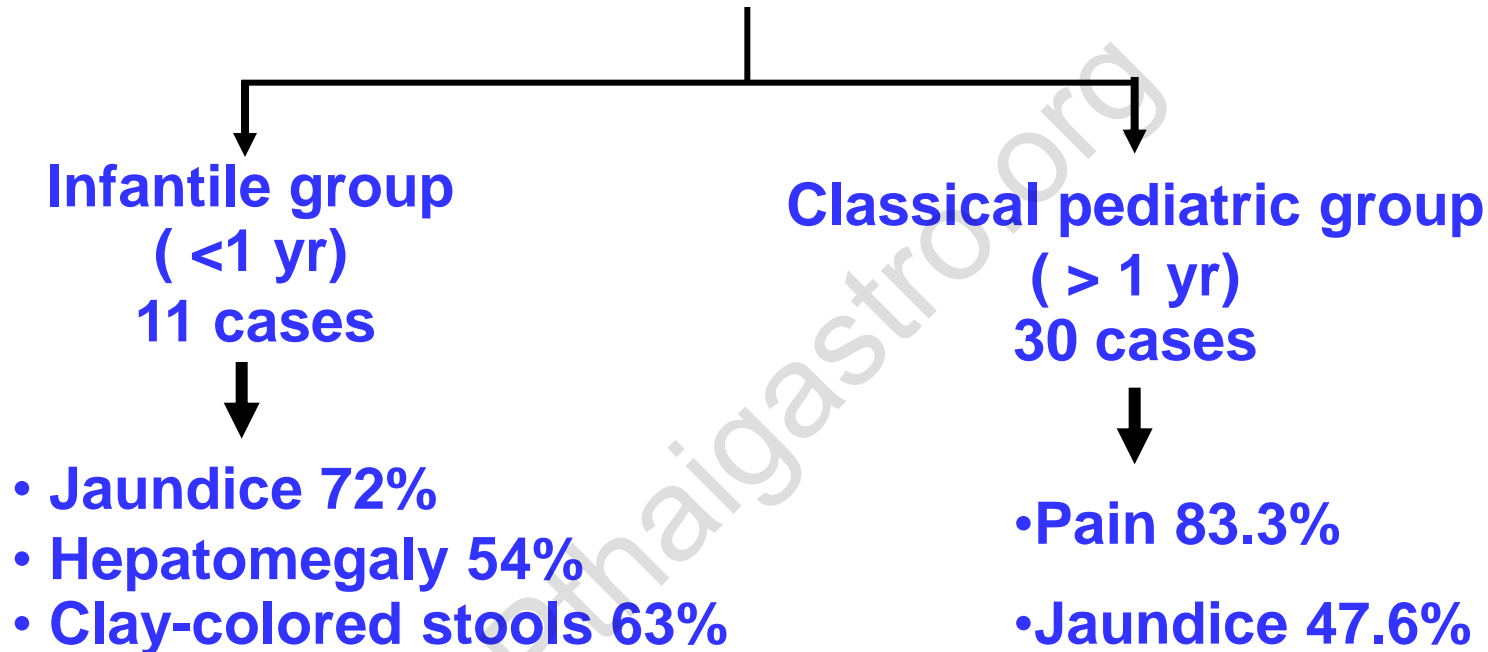
Clinical features

- **May be diagnosed prenatally on ultrasound**
 - **25% present as neonates with prolonged jaundice and cholestasis**
 - **75% later in childhood with triad of:**
 - **Abdominal pain**
 - **Abdominal mass**
 - **Intermittent jaundice**
 - **Occur less than 20%**
- Symptoms asso. With CCs are usually due to asso.complications of ascending cholangitis and pancreatitis**

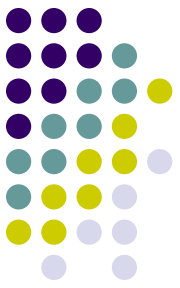


Choledochal cysts in infancy and childhood

41 cases of CC (20 days-11years)



Choledochal cysts in infancy and childhood



- 3 cases had biliary perforation with localized or generalized biliary peritonitis
- Operative management – primary complete excision of cyst with Roux-en-y hepaticodochojejunostomy (n=32)
- 2 postoperative deaths (cirrhotic liver with PHT, deep icterus with poor general condition)

Choledochal Cyst Disease in Children and Adults: A 30-year Single-Institute Experience

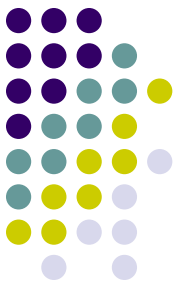


Demographic Information and Medical Comorbidities of 92 Patients with CC

Demographic characteristic	n	%
Female	83	90
Male	9	10
Adults (≥ 16 y)	73	79
Children	19	21

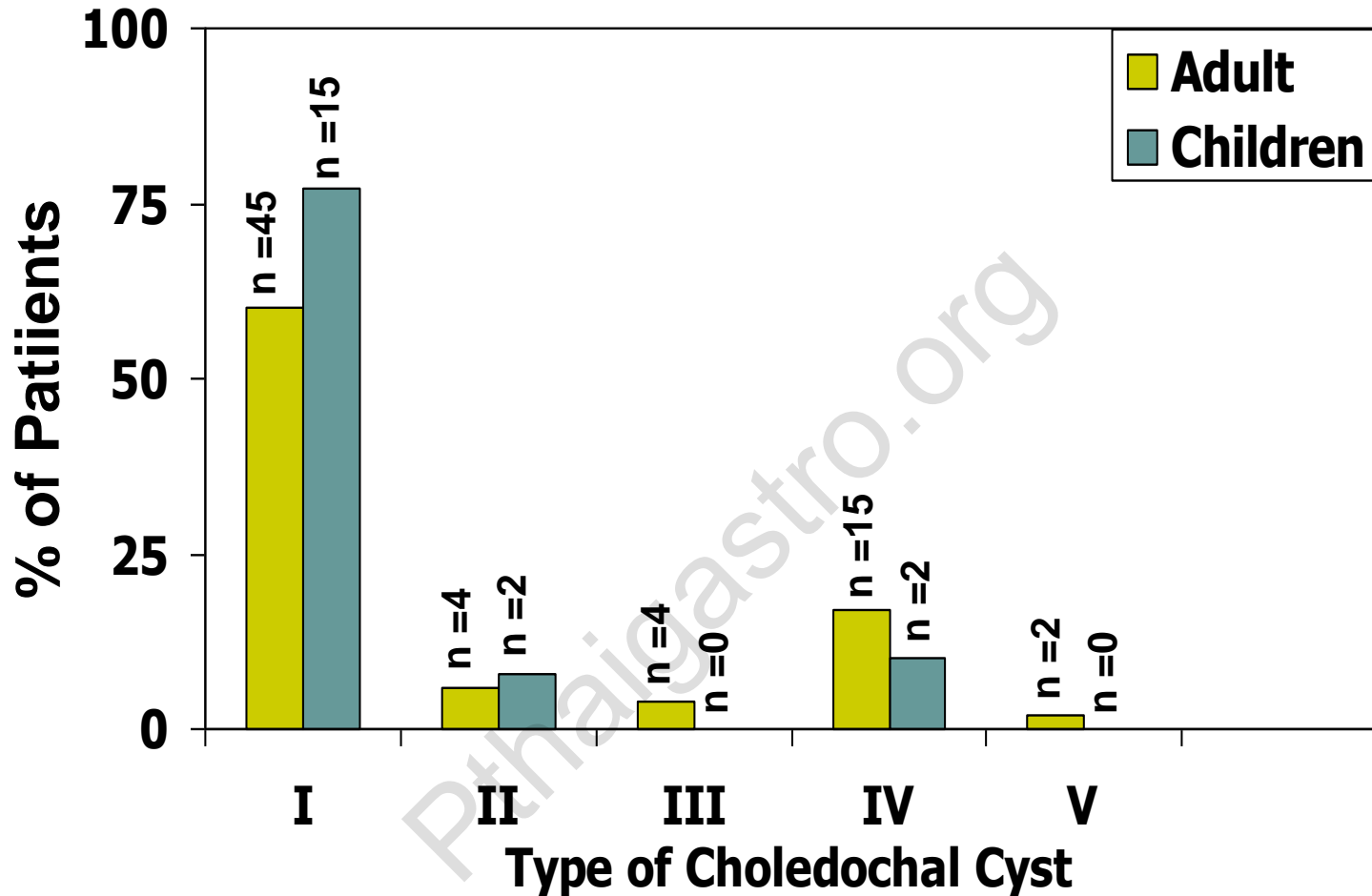
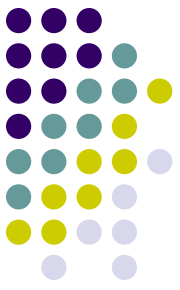
Edil B H,et al. J Am Coll Surg.2008;206:1000-1008.

Choledochal Cyst Disease in Children and Adults: A 30-year Single-Institute Experience



Comparative Presentation Between Children and Adults

Symptom	Overall,%	Child,%	Adults,%	pValue
RUQ pain	91	63	97	<0.001
Jaundice	34	71	25	0.001
Fever/chills	26	25	26	1.000
Pancreatitis	31	31	30	1.000
Cholelithiasis	58	41	62	0.17
Weight loss	8	6	9	1.000
Pruritis	1	0	1	1.000
Nausea/vomiting	47	53	45	0.6
GI bleeding	1.2	0	1.5	1.000



Relative breakdown of choledochal cyst type with respect to adults and children in 89 patients

Comparative Postoperative Morbidity Between Adults and Children



Morbidity	Total %	Child %	Adult %	P Value
Overall complication	34	6	41.0	0.008
Reoperation(n=2)	2	0	3	1.000
Pancreatitis (n=4)	5	0	6	0.581
Cardiac(n=1)	1	0	1	1.000
Pneumonia (n=)	1	6	0	0.198
Wound infection (n=5)	7	0	9	0.594
Intraabdominal abscess(n=5)	7	0	9	0.594
Cholangitis (n=6)	8	0	10	0.336
Bile leak (n=5)	7	0	9	0.594

Characteristic of Five Patients with Malignant disease at Presentation in Association with Their Choledochal Cysts

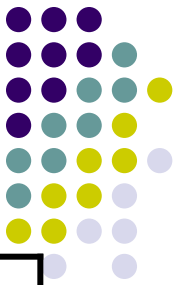


Age, y	Symptoms	Operative intervention	Type of cyst	Pathology	Tumor size, cm	Lymph node involvement	Margin status	Outcomes
3	Jaundice	Extrahepatic biliary excision with RYN HJ	I	Embryonal rhabdomyosarcoma	NA	No	+	Alive, 11y
34	Abdominal pain, 25-lb weight loss	Pancreatico duodenectomy	I	Cholangio carcinoma	2.5	Yes	-	Dead 10.0 mo.
44	Abdominal pain	Hepatic resection, cyst excision, RYN HJ	I	Gall bladder cancer	0.6	No	-	Alive, 6y
45	Abdominal pain, jaundice, F/C, pancreatitis	Pancreatico duodenectomy	I	Cholangio carcinoma	5.0	No	-	Alive, 2y
24	Abdominal pain, jaundice	Extrahepatic biliary excision with RYN HJ	IV	Cholangio carcinoma	0.6	No	+	Dead, 7 mo

Characteristics of patients who did not have malignancy at the time of their cyst excision but who subsequently died

Age, y (gender)	Symptoms	Operative intervention	Type of cyst	Cause of death	Survival time, y
28 (M)	Abdominal pain, F/C	Extrahepatic biliary excision with RYN HJ	IV	Cholangiocarcinoma	8
42 (F)	Abdominal pain,	Extrahepatic biliary excision with RYN HJ	I	Pancreatic adenocarcinoma	21
63 (F)	Abdominal pain, jaundice F/C	Extrahepatic biliary excision with RYN HJ	V	Cholangiocarcinoma	9
68 (F)	Abdominal pain, F/C	Extrahepatic biliary excision with RYN HJ	I	Unknown	14
16 (F)	Abdominal pain,	Extrahepatic biliary excision with RYN HJ	I	Unknown	14

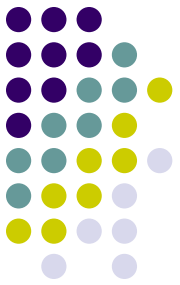
Symptomatology



Symptomatology	Number of patients	%
1. Jaundice	49	66.2
2. Abdominal pain	36	48.7
3. Abdominal mass	31	41.9
4. Acholic stool	24	32.4
5. Vomiting	24	32.4
6. Fever	20	27.0
7. Ascites	4	5.4

Watanatittan S, Niramis R. Choledochal cyst : Review of 74 Pediatric Cases. J Med Assoc Thai 1994.

Amylase level in CDC content



30 cases has amylase level

17 cases amylase level higher than 500 U/dl

16/18 cases age more than 1 yr.
amylase level higher than 500 U/dl

1/12 cases age less than 1 yr.
amylase level higher than 500 U/dl

Types of choledochal cyst



Type	Number of patients
Type Ia	38
Ib	0
Ic	6
Type II	0
Type III	0
Type IV A	29
Type IV B	0
Type V	1
Total	74

Watanatittan S, Niramis R. Choledochal cyst : Review of 74 Pediatric Cases. J Med Assoc Thai 1994.

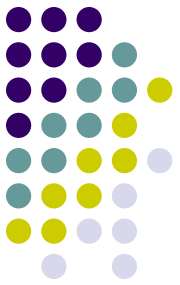


Frequency of Symptoms and signs associated with choledochal cysts in 17 adults

Symptoms and signs	Frequency (%)
Jaundice	70.6
Abdominal pain	58.8
Fever	52.9
Abdominal mass	41.2
Classical triad	10.8

Akaraviputh T et al. Surgical management of adult choledochal cysts. J Med Assoc Thai 2005.

Hepatobiliary disease associated with choledochal cysts in 17 adults



Pathologic finding	No. of patient (%)
Biliarylithiasis	8(47.0%)
Acute pancreatitis	3(17.6%)
Cholangiocarcinoma	1(5.9%)

Akaraviputh T et al. Surgical management of adult choledochal cysts. J Med Assoc Thai 2005.

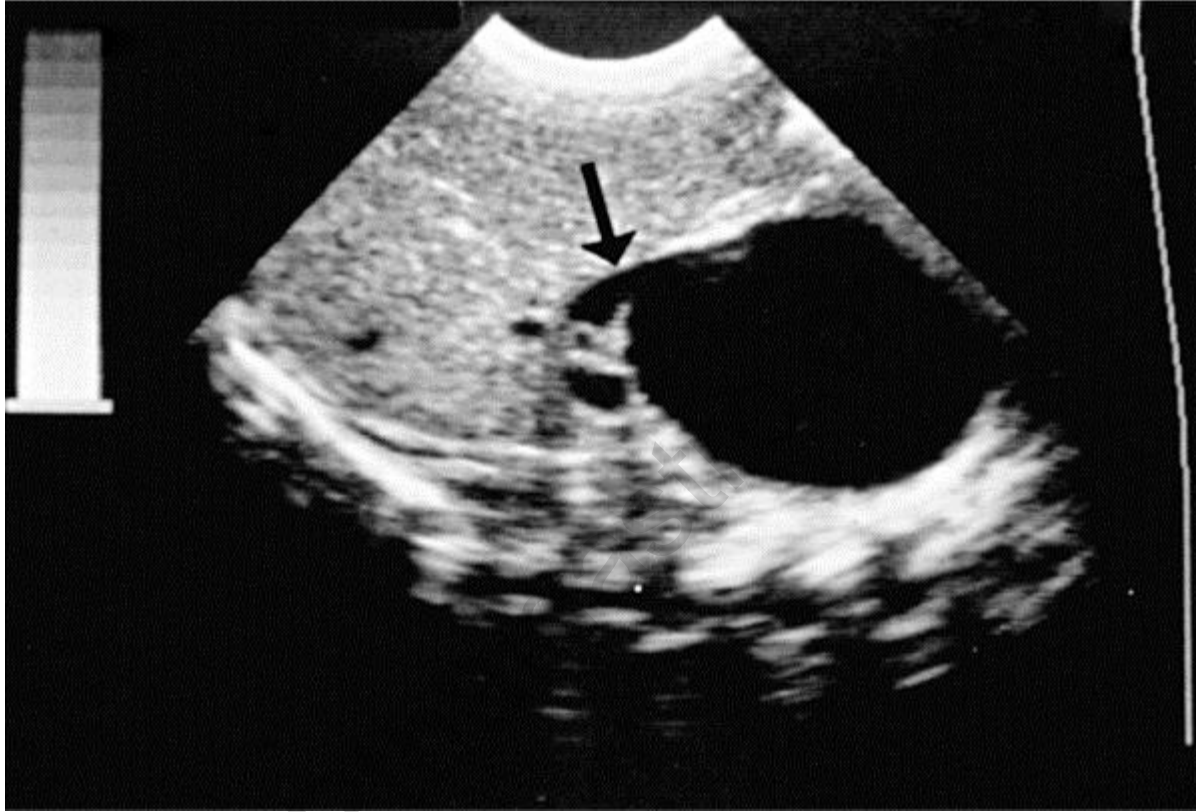
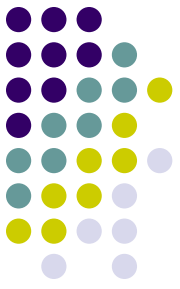
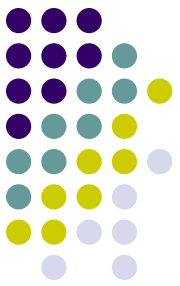


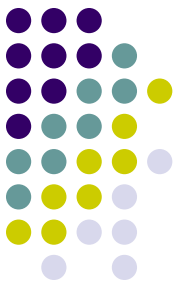
Figure 59-7 Ultrasonographic demonstration of a type I choledochal cyst in an infant with cholestasis. A large cystic mass in the right upper quadrant is shown on this transverse scan. The point of juncture with the common bile duct is delineated by the *arrow*.



Pathogenesis

- **Babbitt's theory : abnormal pancreaticobiliary duct junction (APBDJ) allows mixing of pancreatic and biliary juices, which then activates pancreatic enz. (50-80% of CCs are associated with APBDJ)**
- **Purely congenital in nature, with distal obstruction due to aganglionosis and proximal dilatation (similar to Hirschprung disease)**
- **Associated with many different developmental anomalies**

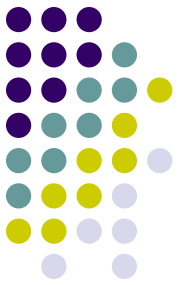
Hepatobiliary pathology associated with the choledochal cysts



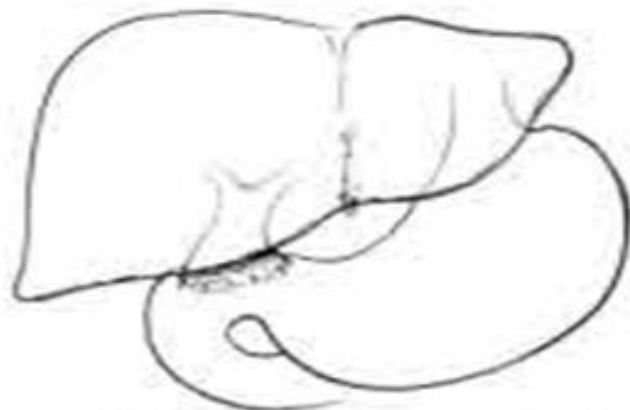
Pathology	Number	Percentage
Anomalous pancreato-biliary junction	10	63
Primary ductal stricture	2	13
Secondary ductal stricture	5	31
Aberrant hepatic artery	4	25
Prominent accessory papilla	1	6
Accessory gallbladder	1	6
Cholelithiasis	7	44
Cystolithiasis	6	38
Intrahepatic stones	2	13

Treatment

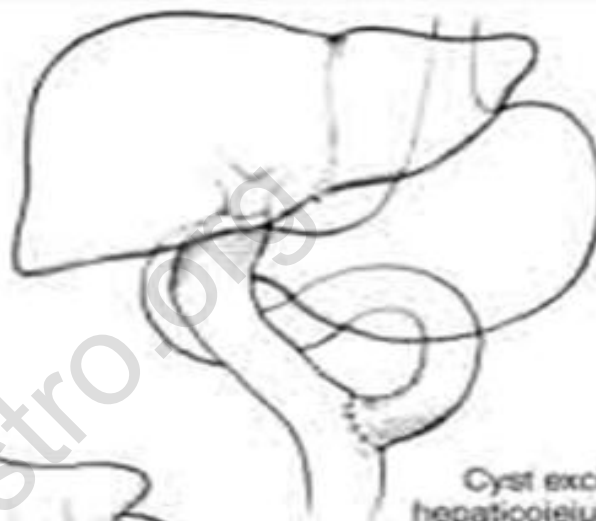
- **Cyst excision and Roux-en-Y hepaticojejunostomy**



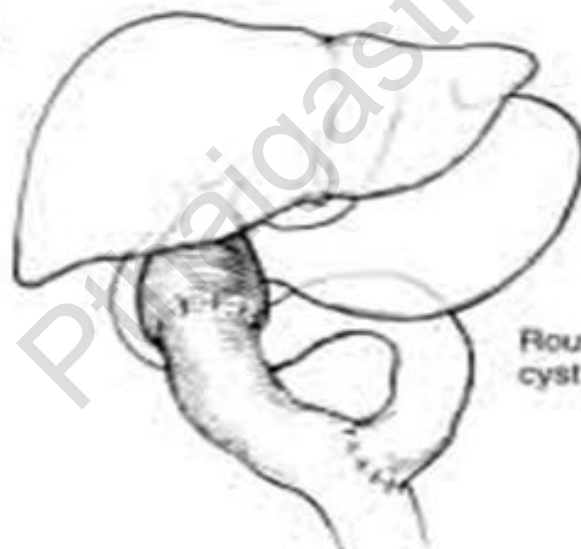
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Cyst duodenostomy



Cyst excision
hepaticojejunostomy

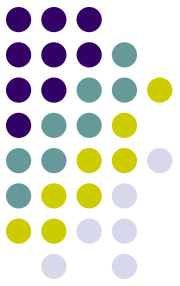


Roux-en-Y
cyst jejunostomy

Ppt.aigastro.org

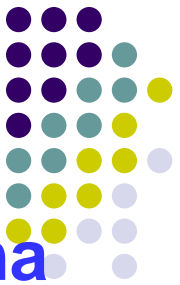
Complication

- **Cholangitis**
- **Pancreatitis**
- **Biliary cirrhosis**

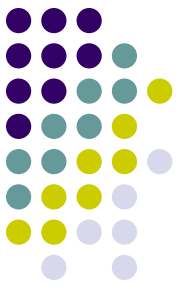


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Malignancy



- Adenocarcinoma 73-84%, anaplastic carcinoma 10%, undifferentiated cancer 5-7%, squamous cell carcinoma 5% and other carcinoma 1.5%
- Site of cancer : extrahepatic bile duct 50-62%, gallbladder 38-46%, intrahepatic duct 2.5%, liver and pancreas 0.7%
- 68% ass. With type-I, 5% type-II, 1.6% type-III, 21% type-IV, and 6% type-V CCs
- Abnormal pancreaticobiliary duct junction has 16-55% risk of malignancy with or without bile duct dilation



Malignancy

- **Cancer occurs as a result of chronic inflammation, cell regeneration and DNA breaks, leading to dysplasia**
- **Recurrent cholangitis or pancreaticobiliary reflux**
- **Chronic postobstructive infection by gram-negative bacteria metabolizes bile duct into carcinogens**

Singham J et al. Review Choledochal cysts. Can J surg 2009.