

IT IS NOT EASY BEING GREEN



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CONGENITAL ANOMALIES

- Infrequent
- Agenesis
- Duplication
- Ectopic gallbladder
- Congenital Stenosis of Cystic Duct

NEONATAL CONDITIONS



■ Acute Gallbladder Hydrops

- Can occur in neonates or older children
- Edema around gallbladder and CBD
- Palpable mass (neonates)
- Fever, pain, mass (older children)
- Neonates- Sepsis cystic duct stenosis or agenesis
- Older children- Sepsis, dehydration,, scarlet fever, familial mediterranean fever leptospirosis, Kawasaki
- Treatment- Expectant, antibiotics, ,early feedings
- F/U- Serial US
- Clinical deterioration, worsening pain - cholecystectomy



BILE ASCITES

- Idiopathic perforation of bile ducts
- Progressive abdominal distention and jaundice
- May be associated with episode of sepsis or ABO incompatibility
- Almost universally perforation at junction of cystic duct and CBD
- At operation cholangiogram through GB
- Perforation seals with drainage
- Aggressive repair not indicated



BILE SLUDGE SYNDROME

- **Extrahepatic obstruction of bile ducts by biliary sludge in infants without anatomic abnormalities**
 - Bile in neonates tends to be highly saturated in cholesterol due to decreased bile salt concentration
 - Inefficient bile acid uptake
 - Impaired bile acid conjugation
 - Decreased synthesis and decreased excretion.
 - Prolonged fasting, TPN, blood transfusions (hemolysis), dehydration, gallbladder stasis, CF, chronic furosemide therapy, SBS, ileal resection for NEC are predisposing factors
 - Treatment- Enteral feeds, Actigall, CCK
 - Spontaneous resolution has been reported.



NEONATAL CHOLELITHIASIS

- 44% have documented history of bile sludge syndrome
- 43% of children on long-term TPN will develop gallstones
- Same risk factors as bile sludge syndrome
- Phototherapy (in the absence of TPN)
- Spontaneous resolution on serial US in 20% (2-20 days after cessation of TPN)
- If stones are calcified they will not resolve
- Non- operative treatment for one year is reasonable
- Laparoscopic cholecystectomy for symptomatic patients and those with calcified stones.



TPN

- 43% of children on long-term TPN will develop gallstones
- Mechanism unclear
- Lack of enteral feeds leads to lack of GB contraction and impaired enterohepatic circulation of bile salts
- Bile composition changed by infused AA
- Administration of fat in the TPN ameliorates the effect of infused AA
- Composition of stones in children <10y is different (more calcium carbonate and black pigment stones)

NEONATAL CHOLEDOCOLITHIASIS



- Spontaneous resolution reported in some patients
- Usually requires operative intervention
- ERCP and sphincterotomy
- Open irrigation through cystic duct vs. trans-duodenal sphincterotomy and sphincteroplasty
- If bile duct perforation present-peritoneal irrigation and drainage.



HEMOLYTIC CHOLELITHIASIS

- Age dependent
- Pigment stones
- Sickle cell disease (50% gallstones by 20 years of age)
 - No difference in bile of stone formers and non-stone formers
 - Stone formers- larger fasting and post-prandial gallbladder volumes
 - Stasis important contributing factor leads to sludge formation (cholecystectomy indicated, approx 65% will develop stones)
 - Due to potential for complications cholecystectomy indicated for asymptomatic cholelithiasis



HEMOLYTIC CHOLELITHIASIS

- **Sickle Cell Disease (cont.)**
 - Highest risk of postoperative complications:
 - Sickle cell events 19%, PACU events 11%, complications 10%, postoperative surgical events 4%
 - Death 1%
 - Acute chest syndrome can be seen in up to 20% of patients undergoing abdominal surgery



HEMOLYTIC CHOLELITHIASIS

- **Sickle Cell disease (cont)**
 - Incidence of sickle cell events may be higher in patients not transfused
 - Attention to hydration, Hb > 10g/dl, oxygenation and antibiotics (many are autosplenectomized) in perioperative period



HEMOLYTIC CHOLELITHIASIS

- **Hereditary Spherocytosis**
 - Incidence-43-63%
 - US in preparation for splenectomy
 - Concurrent laparoscopic splenectomy and cholecystectomy when possible
- **Thalassemia major**
 - Incidence: 6% of children 6-10y
 - Up to 45% in children 11-14 y
 - Hypertransfusion has decreased the incidence due to bone marrow suppression.



CHOLESTEROL CHOLELITHIASIS

- Solubility of cholesterol depends on the concentration of lecithin, bile salts and cholesterol within bile
- Stones result when bile cannot solubilize all of the cholesterol
- Any disturbance in any of these three substances may render bile lithogenic
- Although risk factors have been described, in most patients no predisposing factor can be identified

CHOLESTEROL CHOLELITHIASIS



- Incidence increasing and surpassing hemolytic diseases
- Typical patient obese young girl (F:M 11-22:1 strong influence of estrogens and progesterone)
- Risk factors-Pregnant adolescents, positive family history, oral contraceptives, CF, chemotherapy (Wilms, NB Hodgkins)
- Pain, fatty food intolerance
- Can occur in infants after TPN, ileal resection
- Dx-US
- Tx- Laparoscopic cholecystectomy



COMPLICATIONS OF CHOLELITHIASIS

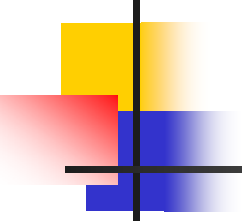
- Pain, weight loss
- Acute cholecystitis
- Choledocolithiasis
- Cholangitis
- Biliary pancreatitis



BACTERIOLOGY

- Bile is normally sterile
- Presence of positive cultures influenced by biliary disease and patient's age
- 11%-30% of patients with symptomatic gallstones have positive bactibilia
- *Escherichia coli* and *Klebsiella* most common G. negative species isolated
- *Bacteroides* and *Clostridium* play a small but significant role in biliary infections
- Polimicrobial infections are more common in patients with cholangitis

ACALCULOUS CHOLECYSTITIS




- Can appear after patient has been resuscitated from primary sepsis, trauma, burns, or shock
- Hydrops of the GB becomes secondarily infected
- Deterioration and signs of sepsis in a previously stable PICU patient
- Dx- US- Distended GB , edema of the wall, echogenic debris
- Mild cases –Medical management with Abx
- Failure to respond to Abx- Cholecystectomy
- Very ill patients- Cholecystostomy, percutaneous or open



BILIARY DYSKINESIA

- Distinct clinical entity occurring in older children
- Poor contractility of gallbladder
- Cholesterol crystals found in GB bile
- Association with chronic cholecystitis
- Nausea, RUQ pain associated with meals
- Tx- Laparoscopic cholecystectomy

**TABLE 105-2 Patients Requiring
Laparoscopic Cholecystectomy at Children's
Mercy Hospital, Kansas City, MO, 2000-2003**

Total Patients	93
Male	31
Female	62
Mean age (yr)	12.3 (2-20)
Mean weight (kg)	55.3 (11-126)
Hematologic disease	
Sickle cell	9
Hereditary spherocytosis	6
Pyruvate kinase deficiency	1
Biliary dyskinesia	3
Acute symptomatic conditions	
Acute biliary colic	6
Gallbladder pancreatitis	2
Jaundice	1



BILIARY DYSKINESIA

Biliary dyskinesia:

Defined as ultrasonography without evidence of cholelithiasis with clinical symptoms of biliary colic

HIDA scan: performed in dynamic fashion over 75 minutes with injection 1 mg of CCK at 45 minutes. EF <35% considered abnormal

RESULTS

- 184 patients underwent Lap Chole
 - May 2003- May 2006
 - 117 for biliary dyskinesia with 108 available for follow up
 - 48 for cholelithiasis
 - 17 for chronic cholecystitis
 - 2 for pancreatitis
 - Mean age 14.1 years old



RESULTS

- Of those undergoing laparoscopic cholecystectomy for biliary dyskinesia

...97.1% claimed surgery beneficial

- 63.9% reported resolution of symptoms
- 27.8% reported improvement in symptoms
- 8.3% reported failure of intervention



RESULTS

- Presenting symptoms and Outcomes

Symptoms	Post cholecystectomy outcomes			(n)
	Resolved	Improved	Failure	
Nonspecific abdominal pain	70.4%	22.2%	7.4%	27
Biliary colic	61.7%	29.6%	8.6%	81



RESULTS

- Scintigraphy and Postoperative Outcomes:

EF on HIDA	Post cholecystectomy outcomes			(n)
	Resolved	Improved	Failure	
EF <15%	67.7%	23.1%	9.2%	65
EF 15-35%	56.8%	40.5%	0.7%	37
EF >35%	60%	20%	20%	5

$p > 0.5$



RESULTS

- **Complications: 3/184**
 - Wound infection = 1
 - Postoperative ileus = 1
 - Incisional hernia = 1

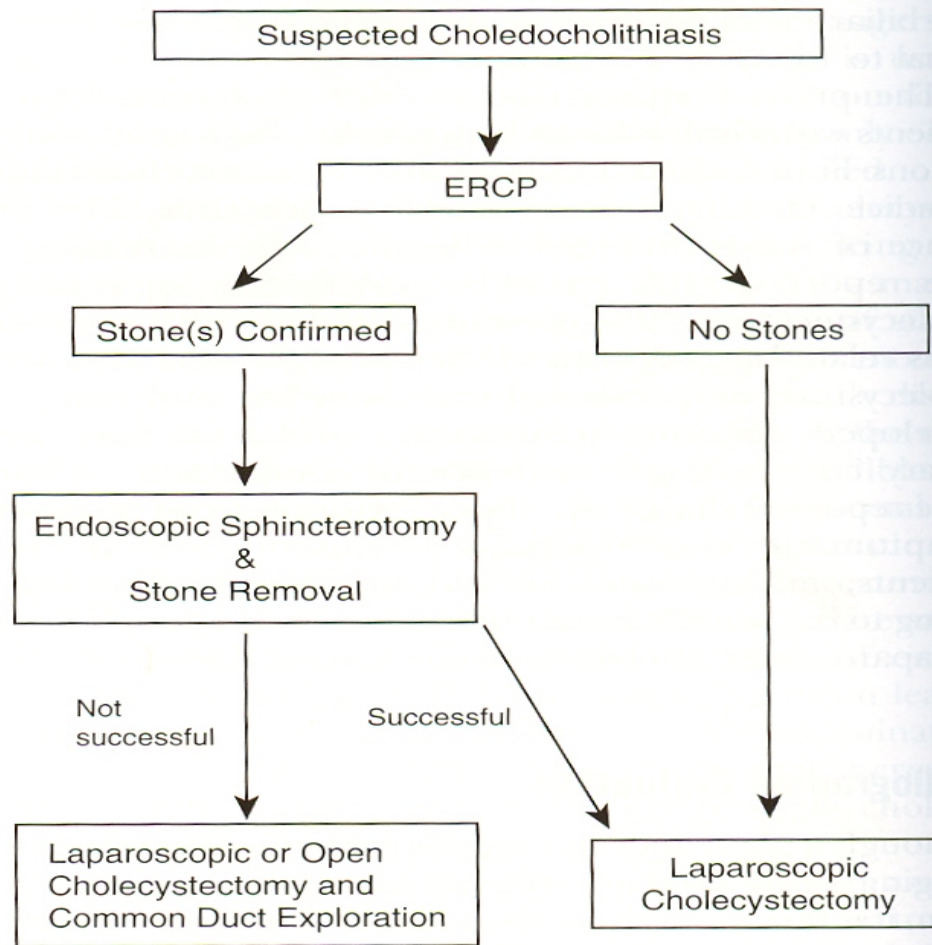
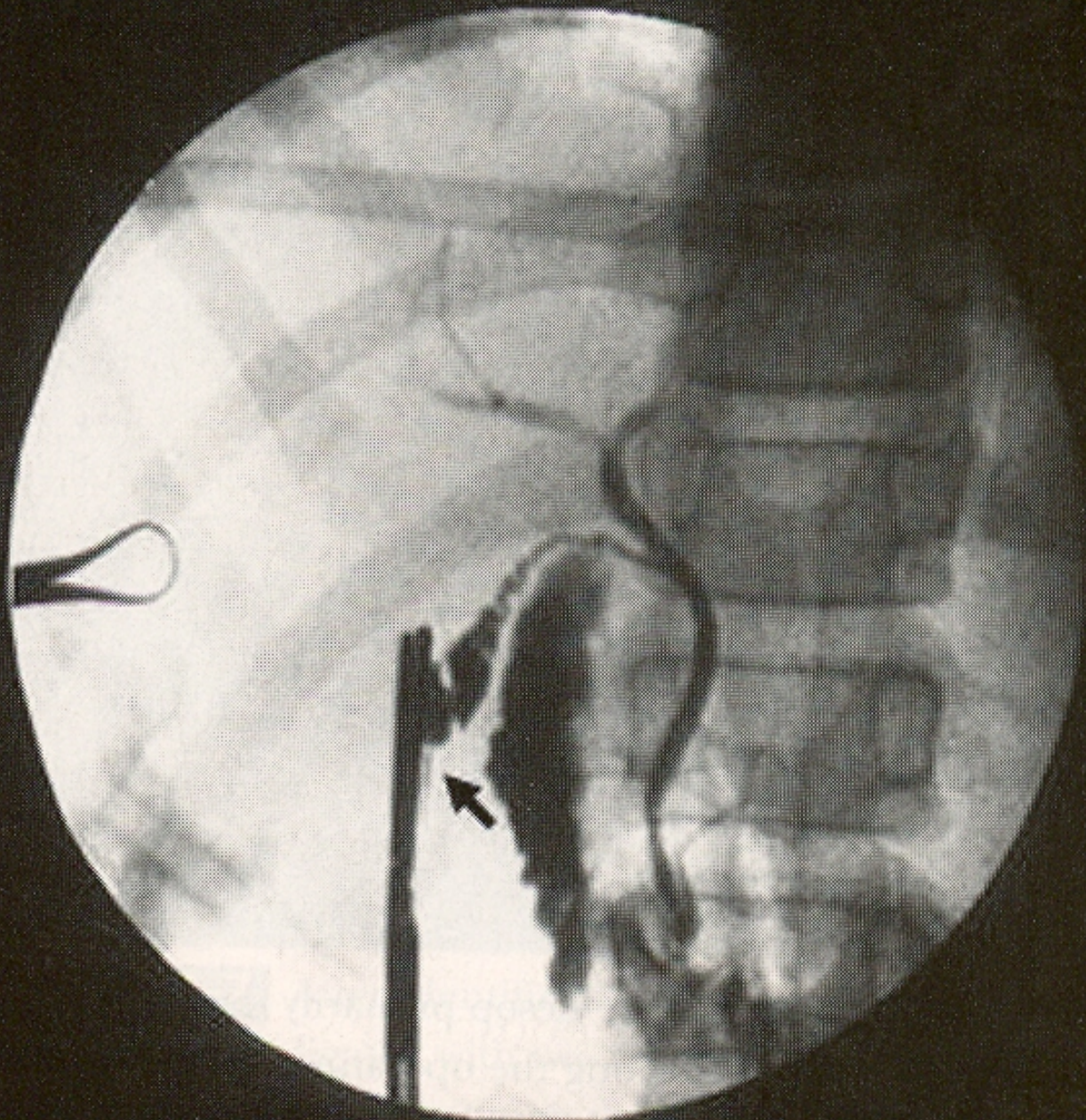


Figure 105-1 This algorithm depicts the authors' preferred strategy for managing patients with suspected choledocholithiasis. Preoperative ERCP is favored with stone extraction and sphincterotomy, if necessary, before the laparoscopic cholecystectomy. If the ERCP does not show evidence of choledocholithiasis, then laparoscopic cholecystectomy is performed. The reason this approach is favored is that, at the time of the laparoscopic cholecystectomy, the surgeon knows whether a laparoscopic or open choledochal exploration is needed.



INTRAOPERATIVE CHOLANGIOGRAM

- Indications:
 - Obstruction
 - Elevated alkaline phosphatase
 - Dilated CBD
 - Jaundice
 - Multiple small stones
 - Pancreatitis
 - Unclear anatomy at operation



ny in infants and small children is depicted. Note the clamp across the



CHOLEDOCOLITHIASIS

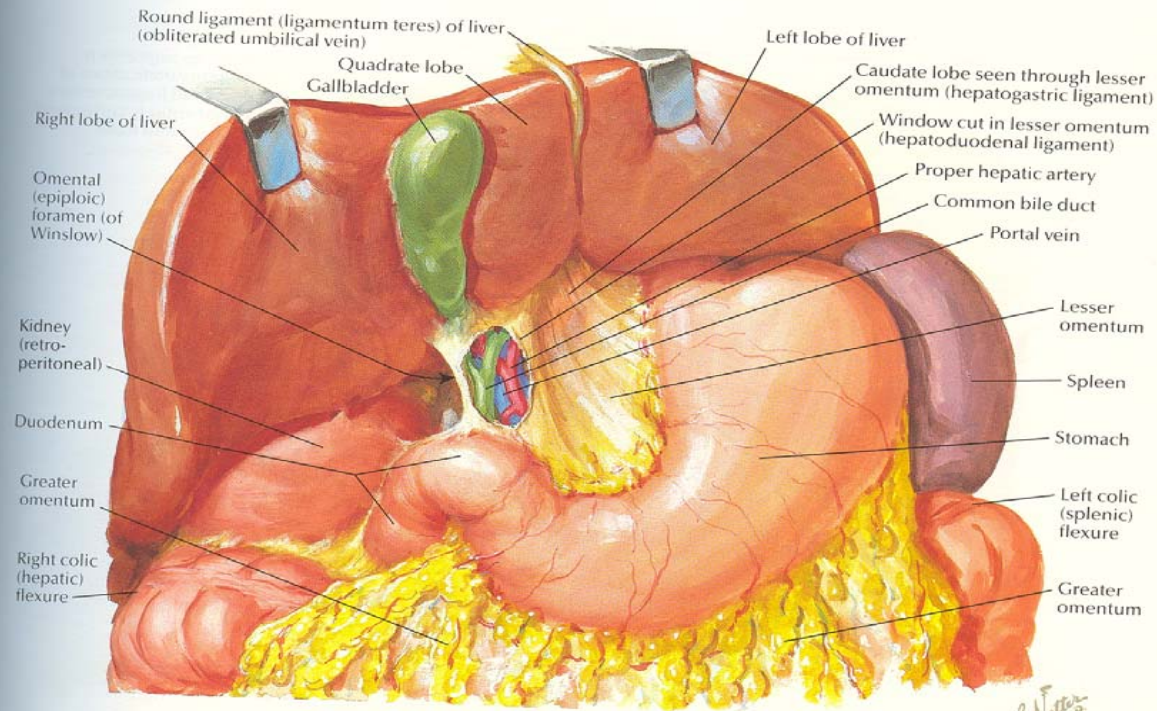
- **THERAPEUTIC OPTIONS:**
 - ERCP
 - Open CBD exploration with T tube
 - Laparoscopic CBD exploration (choledocoscopy)
 - When?



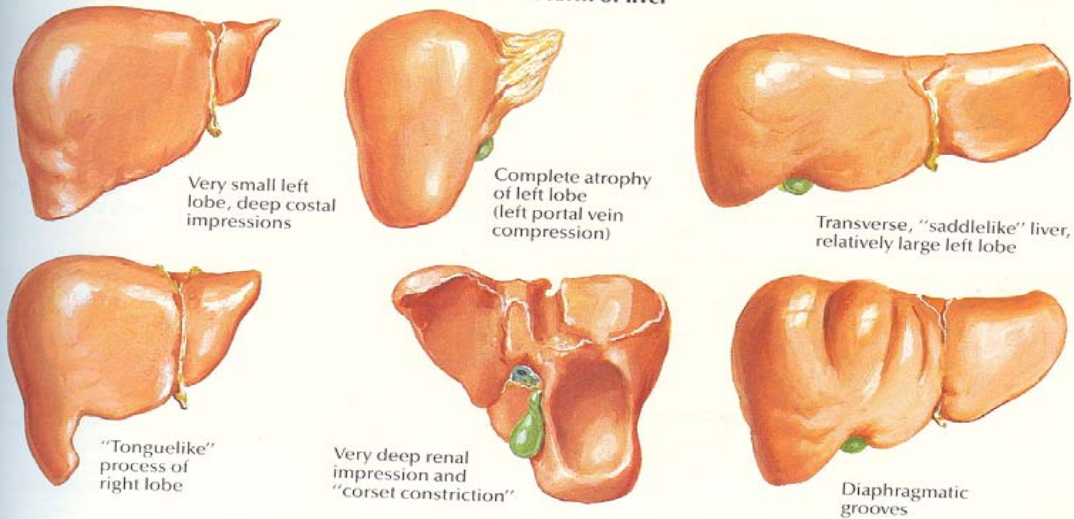
COMPLICATIONS

- Conversion to open- 2%
- Infection
- Bleeding
- Bile leak- 1.5%-2%
- Bile duct injury- 0.1%-0.2%

Liver In Situ and Variations in Form

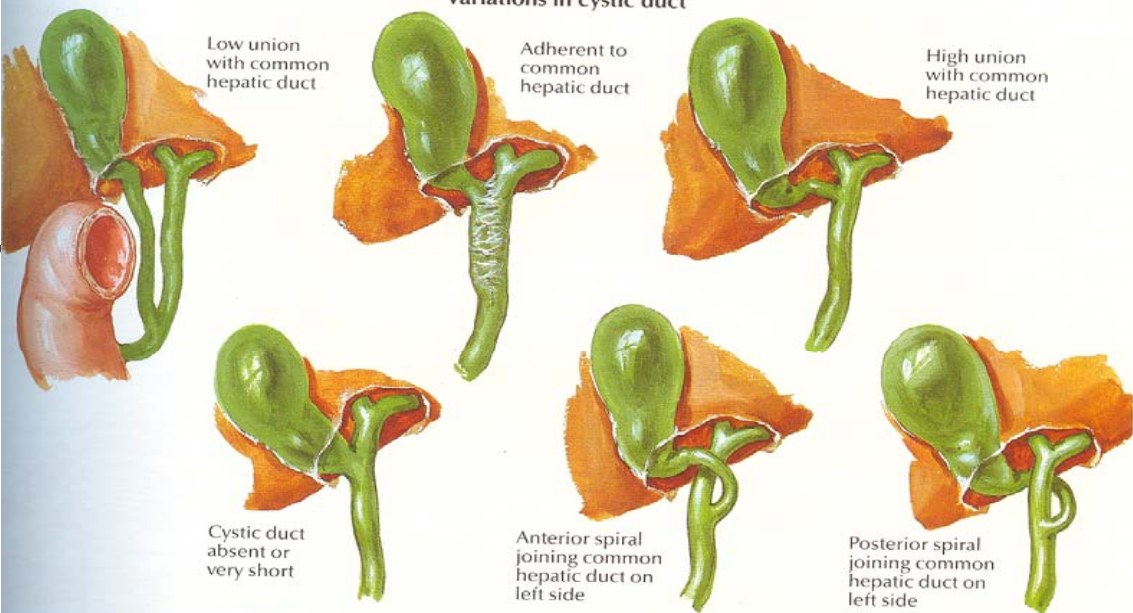


Variations in form of liver

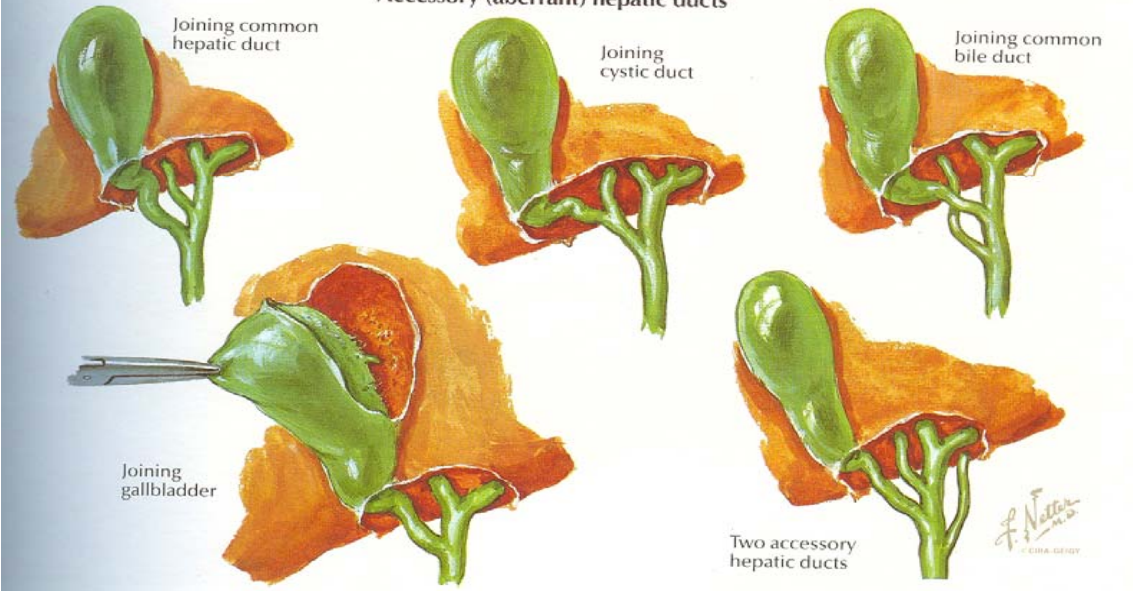


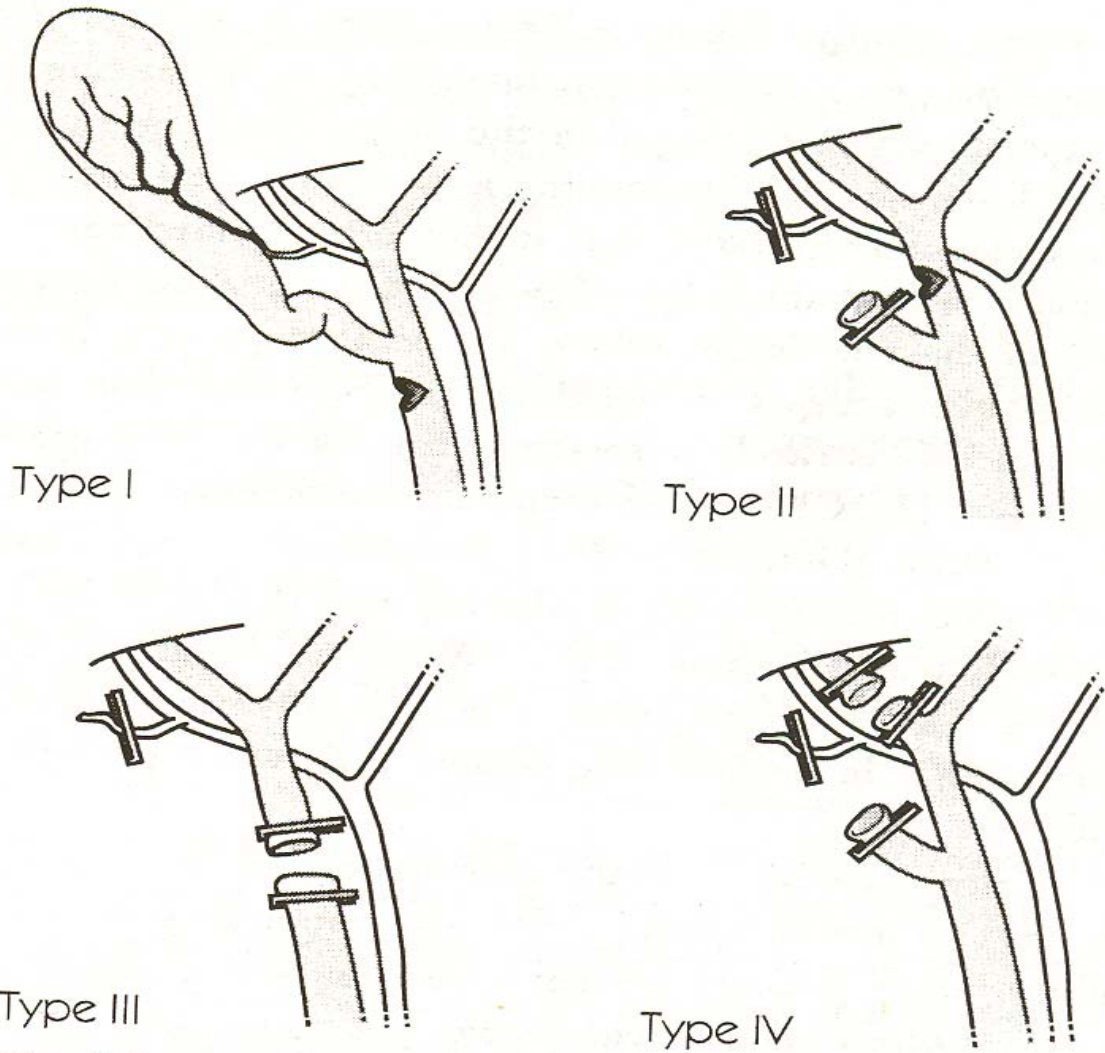
Variations in Cystic and Hepatic Ducts

Variations in cystic duct



Accessory (aberrant) hepatic ducts





Type I

Type II

Type III

Type IV

Fig. 8-2. Classification scheme for bile duct injuries (see text).



CONCLUSION

Laparoscopic cholecystectomy is safe, efficacious and durable in children suffering from biliary dyskinesia and a diverse number of diseases of the gallbladder in the pediatric age groups.