



# **PEDIATRIC SURGERY Update** ©

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### **Frantz Tumor**

Papillary cystic tumor of the pancreas, also known as Frantz tumor (FT) since 1959, occurs predominantly in girls and young women (mean age 21 years). Abdominal pain and a slowly growing incidentally found epigastric mass is the most common complaints, associated at times with weight loss, anorexia and vomiting. FT is well-encapsulated, shows solid and hemorrhagic patterns, contain PAS-positive cytoplasmic or prozymogen granules as seen in acinar cell tumors and behaves as a low-grade malignancy. CT scans suggest the diagnosis (thick capsule, calcifications, mixed solid and cystic patterns, grows toward the outside of the pancreas). Differential diagnosis includes traumatic pseudocysts, serous and mucinous cystadenomas of the pancreas. Immunohistochemically the tumor is positive for alpha-1-antitrypsin while negative for insulin and glucagon. Complete removal is the treatment of choice for tumor arising in any part of the pancreas. FT is frequently amenable to local resection and has a good long-term survival rate after excision. Metastasis (liver) or local recurrence occurs in 10% of cases. Older age at diagnosis or recurrence disease increases the malignant biological behavior of the tumor. Radiotherapy and, or chemotherapy are of no use for its treatment.

### **References**

- 1- Todani T, Shimada K, Watanabe Y, Toki A, Fujii T: Frantz's tumor: a papillary and cystic tumor of the pancreas in girls. *J Pediatr Surg* 23(2):116-21, 1988
- 2- Tait N, Greenberg ML, Richardson AJ, Osborn RA, Little JM: Frantz's tumour: papillary and cystic carcinoma of the pancreas. *Aust N Z J Surg* 65(4):237-41, 1995
- 3- Horisawa M, Niinomi N, Sato T, Yokoi S, Oda K, Ichikawa M, Hayakawa S: Frantz's tumor (solid and cystic tumor of the pancreas) with liver metastasis: successful treatment and long-term follow-up. *J Pediatr Surg* 30(5):724-6, 1995
- 4- Yang YJ, Chen JS, Chen CJ, Lin PW, Chang KC, Tzeng CC: Papillary cystic tumor of the pancreas in children. *Scand J Gastroenterol* 31(12):1223-7, 1996
- 5- Rivera M, Ortiz VN, Duran N, Trujillo O: Solid and papillary neoplasm of the pancreas: a case presentation. *Bol Asoc Med P R* 90(4-6):91-2, 1998
- 6- Ooi LL, Ho GH, Chew SP, Low CH, Soo KC: Cystic tumours of the pancreas: a diagnostic dilemma. *Aust N Z J Surg* 1998 Dec;68(12):844-6

### **Bile Duct Injury**

Laparoscopic cholecystectomy (LC) has replaced the open procedure as the treatment of choice in gallbladder disorders. With the lap technique an increase two- to threefold in the incidence (0.5%) of bile duct injuries (BDI) has also occurred. Inappropriate anatomy identification and aberrant situations are the most common cause of BDI. BDI can be classified into transection, lacerations, leaks or strictures. Disease complexity, increasing

age, male gender and admission to a teaching hospital are associated with an increase risk of injury. Classic injuries involve clipping & dividing the CBD for the cystic duct. Other BDI may result from stenting injury, thermal injury, lose cystic duct clip or an accessory “Luschka” duct. Such injuries cause two principal clinical manifestations: 1) bile leaks with pain and secondary bile peritonitis, and 2) biliary obstruction due to partial or complete hepatic or common duct ligation (jaundice) or late stricture formation. US, HIDA scan or CT-Scan followed by ERCP or PTC may help diagnosed and define the exact location and nature of the BDI. Complete proximal obstruction may need transhepatic biliary catheters in each radical duct to relieve obstruction. A bile leak may need percutaneous drainage for sepsis control. Cystic or accessory ducts leaks can be dealt with endoscopic ampullary stenting or re-laparoscopic ligation if identified early. The most effective means of reconstructing a major BDI is using roux-en-y hepaticojejunostomy after inflammation and adhesions of the initial insult have subsided. Intraoperative cholangiography appears to protect against BDI by preventing misidentification of anatomy. Conversion to open cholecystectomy is essential in avoiding these injuries when the anatomy remains uncertain.

### References

- 1- Ferguson CM; Rattner DW; Warshaw AL: Bile duct injury in laparoscopic cholecystectomy. *Surg Laparosc Endosc* 2(1):1-7, 1992
- 2- Davidoff AM; Pappas TN; Murray EA; Hilleren DJ, Johnson RD; Baker ME; Newman GE; Cotton PB; Meyers WC: Mechanisms of major biliary injury during laparoscopic cholecystectomy. *Ann Surg* 215(3):196-202, 1992
- 3- Lee VS; Chari RS; Cucchiaro G; Meyers WC: Complications of laparoscopic cholecystectomy. *Am J Surg* 165(4):527-32, 1993
- 4- Branum G; Schmitt C; Baillie J; Suhocki P; Baker M, Davidoff A; Branch S; Chari R; Cucchiaro G; Murray E; et al: Management of major biliary complications after laparoscopic cholecystectomy. *Ann Surg* 217(5):532-40, 1993
- 5- Bergman JJ; van den Brink GR; Rauws EA; de Wit L, Obertop H; Huibregtse K; Tytgat GN; Gouma DJ: Treatment of bile duct lesions after laparoscopic cholecystectomy. *Gut* 38(1):141-7, 1996
- 6- Lillermoe KD; Martin SA; Cameron JL; Yeo CJ, Talamini MA; Kaushal S; Coleman J; Venbrux AC; Savader SJ, Osterman FA; Pitt HA: Major bile duct injuries during laparoscopic cholecystectomy. Follow-up after combined surgical and radiologic management. *Ann Surg* 225(5):459-68, 1997
- 7- MacFadyen Jr BV; Vecchio R; Ricardo AE; Mathis CR: Bile duct injury after laparoscopic cholecystectomy. The United States experience. *Surg Endosc* 12(4):315-21, 1998
- 8- Fletcher DR, Hobbs MS, Tan P, Valinsky LJ, Hockey RL: Complications of cholecystectomy: risks of the laparoscopic approach and protective effects of operative cholangiography: a population-based study. *Ann Surg* 229(4):449-57, 1999

### Septate GB

Congenital anomalies of the gallbladder includes agenesis (congenital absence), duplications, and septation. Finding a Septate gallbladder (GB) is a very rare event that could herald biliary problems. The septum (which contains smooth muscle fibers) usually divides the GB into two chambers. The anomaly in endoderm occurs during the second month of fetal life when the GB lumen develops. Mostly the malformation runs asymptomatic and it is found accidentally without clinical relevance unless the pinpoint communication between the cavities causes stagnation, inflammation or stone formation.

Children present with history of chronic biliary colicky abdominal pain associated with nausea and vomiting. Symptoms are caused by pressure in the small chambers of the GB along with delay emptying. Diagnosis is made using Ultrasound (flat fundus, elongated and dilated GB) couple with biliary scintigraphy (HIDA Scan). Symptomatic children should be managed with laparoscopic cholecystectomy.

#### References

- 1- Esper E, Kaufman DB, Crary GS, Snover DC, Leonard AS: Septate Gallbladder with Cholelithiasis: A Cause of Chronic Abdominal Pain in a 6-Year-Old Child. J Pediatr Surg 27 (2): 1560-1562, 1992
- 2- Doyle TC: Flattened fundus sign of the septate gallbladder. Gastrointest Radiol 9(4):345-7, 1984
- 3- Mrhac L, Zakko S, Ibrahim A: Imaging of septate gallbladder. Clin Nucl Med 24(3):209-10, 1999

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