



PEDIATRIC SURGERY *Update* ©

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Pancreatic Cysts

Pancreatic cystic lesions are usually inflammatory pseudocyst (90%) or neoplastic process (10%). Distinguishing between them is essential for appropriate surgical therapy. Non-inflammatory neoplastic cysts in children are very rare ductal lesions with a spectrum of histologic characteristics and favorable outcomes. Histologically they include retention cysts, lymphoepithelial cysts, papillary cystic tumors, benign serous cystadenoma, mucinous tumors and mucinous cystadenocarcinoma. Most reported cases occur in females during adolescent years. Mode of presentation includes mild upper abdominal pain and palpable mass. Clinical, radiographic and intraoperative frozen section are non-reliable methods in distinguishing the different types of pancreatic cysts. Preoperative cyst fluid obtained by US or CT-guided percutaneous aspiration can be analyzed for viscosity (mucoïd, viscous, serous), chemical (amylase, lipase), tumor markers (CEA, CA 19-9, CA125) and cytology characteristics. High CEA levels (> 25 ng/ml) indicate that the cyst is either malignant or mucinous (pre-malignant) type. Higher levels of CA 19-9 suggest pseudocysts and serous cystadenomas. Very high CA 125 levels appear predictive of malignancy. Viscosity above 1.63 suggests mucinous tumors. Amylase and lipase content should be low in true pancreatic cysts. Cytology analysis is insensitive unless positive for tumor cells. When the nature of the pancreatic cyst cannot be definitively established by the above methods surgical resection is indicated.

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Mucinous Cystadenoma

Mucinous cystadenoma (MCA) of the pancreas is a benign cystic tumor made of columnar mucin-producing epithelium with a pre-malignant potential rarely seen in children. Most cases are identified in teenage and young adult female patients. Abdominal pain and mass effect is the most common symptom when MCA develops. Mucinous cystadenomas grow

to very large sizes, is often multilocular in imaging studies and can be associated with recurrent pancreatitis (due to cyst and duct communication). CT findings might include large cysts with septa, peripheral calcifications and solid intracystic components. Cysts development has an insidious onset not associated to trauma, a high incidence of biliary tract disease or alcoholism. More than 90% of these slow-growing cysts are located in the body and tail of the pancreas. Aspiration of MCA fluid shows low levels of pancreatic enzymes with high CEA, CA 19-9 tumor markers and M1 mucin antigen levels. Cytology evaluation can reveal mucin-containing cells. Due to premalignant potential of developing a cystadenocarcinoma management of MCA should include complete resection. Enucleation and median pancreatectomy carries a high rate of pancreatic fistula formation. Resection is curative.

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Serous Cystadenoma

Serous cystadenoma (SCA) is a benign microcystic tumor of the pancreas composed of cuboidal epithelium very rarely seen in children. Predominantly seen in young females, this cystic tumor is asymptomatic in one-third of the cases. SCA has a microcystic appearance with cysts less than 2 cm in size with loculations most commonly found in the pancreatic body and tail as a single lesion. CT scan shows well-demarcated multilocular cysts with enhancement that might show central calcifications. SCA is associated with another extrapancreatic neoplasm in 20% of cases. Cytology and biochemical analysis of cyst fluid reveal glycogen-rich cells and very low CEA levels (less than five ng/ml). Oligocystic (large cyst) SCA is an even rarer variety that arise in the head of the pancreas usually presenting in infants as a palpable mass needing resection. Should the diagnosis of SCA be well-established conservative management (observation) can be performed with yearly ultrasound study safely. Otherwise, if the nature of the cystic lesion cannot be established, or complications develop (obstructive jaundice, pancreatic duct dilatation or portal hypertension) resection is the next step in management.

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