Choledochal Cysts

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Algorithmic Approach

Five types of CC are described in this classification. Type I CC (80–90% of all CC) is a fusiform dilation of the common bile duct. Type II CC is a true diverticula of the common bile duct (CBD). Type III CC is an intraduodenal dilation of the common channel also known as choledochocele. Type IVA CC (15–20% of all CC) is multiple dilations of the intra- and extrahepatic biliary tree, whereas type IV B CC is multiple extrahepatic dilations only. Type V CC or Caroli's disease is intrahepatic biliary tree dilation only. Symptoms of CC include abdominal pain, jaundice, and often a palpable abdominal mass if presenting at the age of <10 years. About 20% of patients are older than 20 years of age with the most common symptom of abdominal pain. Untreated CC complications include cholangitis, pancreatitis, and obstructive jaundice. Cancer has been associated with all subtypes of biliary cysts but is most commonly found in type I and type IV CC.

A. Vague patient presentation is often misleading and leads to delayed diagnosis. The most common presenting symptom is abdominal pain [1]. Even though these cysts are congenital, they often present in adult years with vague right upper quadrant symptoms, often leading to cholecystectomy for presumed gallbladder disease. Neonates with obstructive jaundice and palpable abdominal mass are usually diagnosed promptly [2].
B. Ultrasound is often obtained to evaluate for right upper quadrant pain or jaundice. An ultrasound finding of a common bile duct dilation >10 mm should alert the physician to investigate for choledochal cyst [2]. Ultrasound findings suggestive of CC should be investigated with magnetic resonance cholangiopancreatography (MRCP), which is considered to be a gold standard of diagnosing all types of CC. Endoscopic retrograde cholangiopancreatography (ERCP) is unnecessary for diagnosis as it is more invasive and increases the risks of cholangitis or pancreatitis [3, 4].
C. The type of CC dictates the surgical management. Type II requires diverticulectomy or simple excision. Type III CC or choledochocele, on the other hand, requires ERCP with sphincterotomy. The risk of malignancy is reported to be very low in both type II and III CC [2].
D. Type I and IVB CC warrant surgery including complete cyst excision and Roux-en-Y hepaticojejunostomy for restoration of biliary continuity. Some surgeons suggest the use of intraoperative frozen sections to rule out dysplasia or malignancy. However, malignancy

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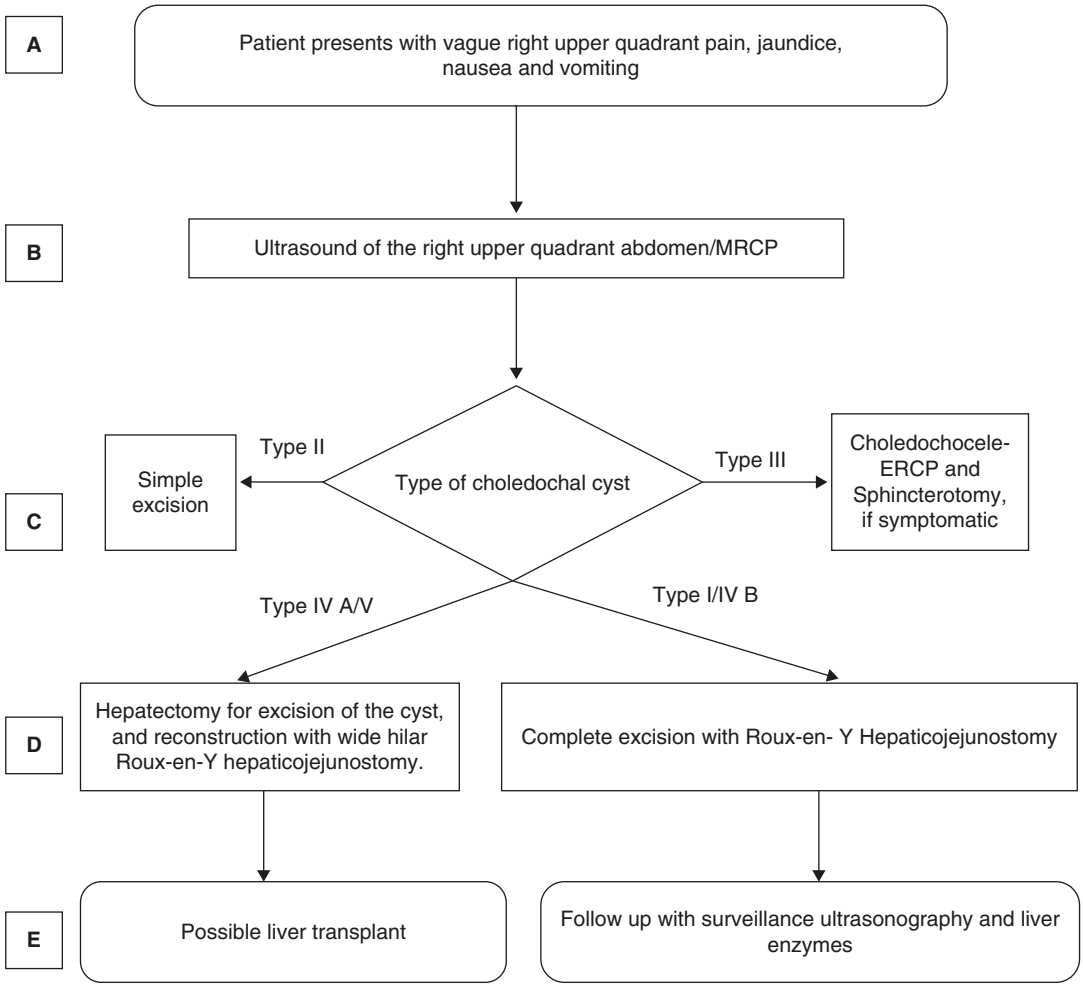
can develop anywhere in the biliary tract including the gallbladder [5]. The literature suggests a 0.7–6% post-excisional malignancy rate in patients with remnant cyst tissue or subclinical malignant disease that is not detected during surgery [2].Type IVA and V (Caroli's disease) CC involve the intrahepatic biliary and may require partial hepatectomy [6, 7].

E. Type V may require liver transplantation for pan-liver involvement. The risk of neoplasia is <7%, but surgical intervention or liver transplant is warranted secondary to cholangitis and liver dysfunction/failure [2]. Given the risk of malignancy in type I and IV CCs, postoperative surveillance is performed with ultrasonography or cross-sectional imaging as well as liver enzymes to detect early cancer. The risk of malignancy is approximately 0.7–6% even in complete excision, primarily due to undetectable cancerous lesions before or at the time of surgery [1, 3, 4].



Algorithm 92.1

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