

Liver Cysts: When and How to Intervene

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Background

Hepatic cysts commonly encountered in daily practice for GI and primary care physicians

Vast majority benign lesions of no clinical significance, but can include malignant and pre-malignant conditions

Prevalence in US estimated between 15-18%

Diagnosis often incidental on imaging for unrelated problems

Estimated only 10-15% patient have symptoms related to cysts

Clin Exp Hepatol 2019; 5,1: 22-29

Differential Diagnosis

Infectious

Hydatid cyst

Amoebic abscess

Pyogenic abscess

Fungal microabscess

Benign

Simple cysts

Polycystic liver disease

Caroli disease

Cavernous hemangioma

Intrahepatic pseudocyst

Traumatic

Biloma

Seroma

Hematoma

Differential Diagnosis

Pre-Malignant

Biliary cystadenoma

Intraductal papillary neoplasm

Caroli disease

Malignant

Cystadenocarcinoma

Cystic HCC

Embryonal sarcoma

Cystic liver metastasis

Simple cysts

Most common liver pathology, affects 2.5-5% population

Slight female predominance

Congenital pathogenesis from bile ducts disconnected from biliary tree

Caroli disease: cavernous ectasia ducts in communication, risk of cancer ~7%

Polycystic liver disease (PCLD): multiple detached, isolated bile ducts; defect in biliary cilia and cholangiocyte proliferation

Simple cysts

Vast majority are asymptomatic, discovered incidentally

Large solitary cysts can present with symptoms:

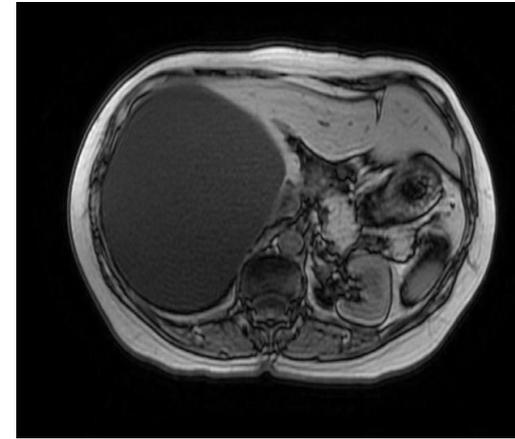
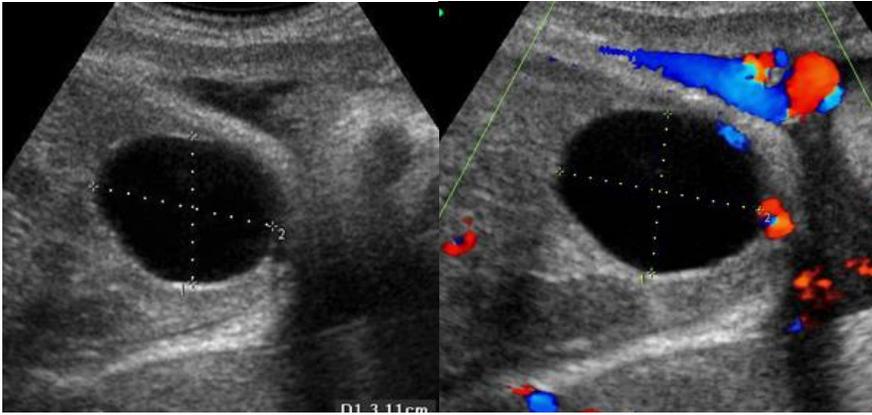
Left: fullness, epigastric pain, early satiety, reflux

Right: Abdominal/flank pain, pain with deep breathing, fullness

Intracystic hemorrhage rare, causes acute onset abdominal pain

Labs typically normal, CA 19-9 elevated – expressed by epithelial lining of cysts

Simple cysts



Anechoic, smooth borders, strong posterior wall echo

Few or no septations

No wall enhancement



Simple cysts

Cyst aspiration

High rate of fluid re-accumulation without sclerotherapy (80%)

Sclerotherapy can be painful

Cyst marsupialization

Laparoscopic or robotic procedure

Removal of cyst wall without transection of liver parenchyma

Symptomatic recurrence reported 10-20%

Liver resection rarely indicated for simple cyst

Clin Exp Hepatol 2019; 5,1: 22-29

Simple cysts

Management of symptomatic cysts

*** Carefully assess attribution of pain to cysts

Minimal to mild symptoms: observation vs percutaneous drain

Moderate to severe: surgical referral to discuss marsupialization

Pts discharged either day of surgery or POD 1

Procedural pain managed under anesthesia

Lowest risk of recurrence

Biliary Cystadenoma

Most common pre-malignant cystic lesion of the liver

Rare, slow growing neoplasm of the bile ducts

Ectopic remnants of embryonic bile ducts

Occurs 1-5/100,000 people

Female:male ration 9:1

Mean age of presentation 45 years

Reported rate of malignant transformation as high as 30%

Biliary Cystadenoma

Typically asymptomatic or non-specific symptoms

Lack of established criteria for pre-operative diagnosis

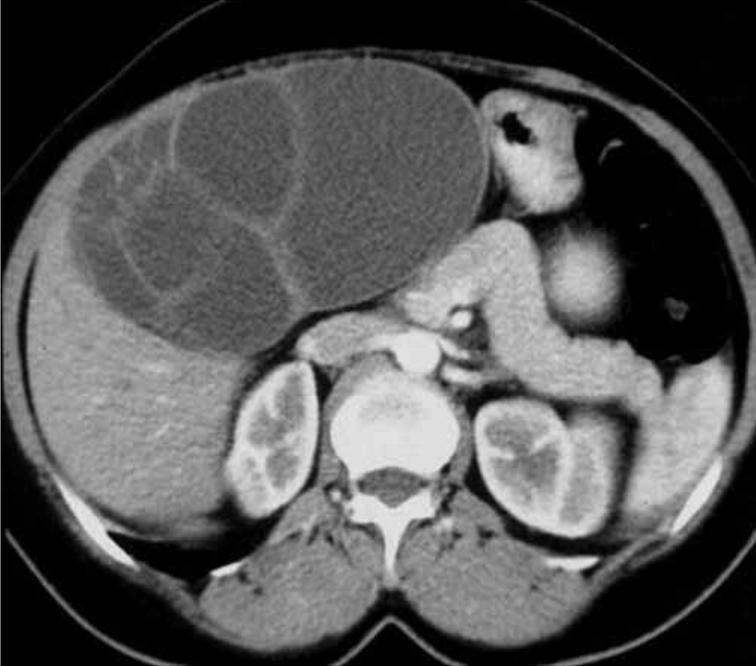
Multi-locular cyst with mucinous fluid divided by irregular thick walls

Classification:

- Mucinous cystic neoplasm contains ovarian stroma

- Intraductal papillary neoplasm communicates with biliary tree

Biliary Cystadenoma



MRI

Multi-locular septated mass

Homogenous low signal on T1

Enhancing wall and septa

FNA or biopsy not recommended for risk of dissemination of tumor

Serum or fluid biomarkers low sensitivity or specificity

Biliary cystadenoma

Liver resection treatment for all cystadenoma due to risk of malignant transformation and inability to rule out adenocarcinoma on imaging

Laparoscopic / robotic approach may be appropriate depending on location

Cyst marsupialization is contra-indication due to the risk of tumor dissemination

** Important to refer to liver surgery specialist if concern for BCA

Biliary cystadenocarcinoma

Very rare cystic malignancy of the liver

Occurs more typically in older, male patients

Elevated liver enzymes/bilirubin more common compared to BCA due to aggressive growth into surrounding liver

Two phenotypes:

Non-invasive: confined to cyst, low recurrence, good prognosis

Invasive: 5-year survival similar to cholangiocarcinoma

Radical liver resection indicated in absence of extrahepatic or metastatic disease

If BCAC is suspected on imaging, staging work up should be performed

Polycystic Liver Disease

Management challenge: wide range of phenotypes / symptoms

Single vs bilobar distribution

Presence or absence of a dominant cyst(s)

Range of symptom severity

Outcomes highly dependent on experience of center

Management dependent on patient's goals

Polycystic Liver Disease

"I want to get rid of my symptoms caused by one or more dominant cysts"

Aspiration Sclerotherapy

Typically lesions >5cm diameter

Reduction of symptoms up to 70% with up to 60% complete relief

Post procedural pain (5-90% reported) or bleeding (2-23%)

J Hepatology 2018; 68 (4):827-837

Polycystic Liver Disease

"I want to get rid of my symptoms caused by one or more dominant cysts"

Fenestration

Multiple cysts can be treated in a single session

Symptom relief in 92% of patients, but 22% symptoms recurred, 24% cyst re-accumulate

Post-op complications include ascites, pleural effusions, bleeding

No RCT comparing Aspiration vs Fenestration

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Polycystic Liver Disease

"I want to get rid of my symptoms caused by one or more dominant cysts"

Resection

No dominant cyst(s) to target, mass effect of liver

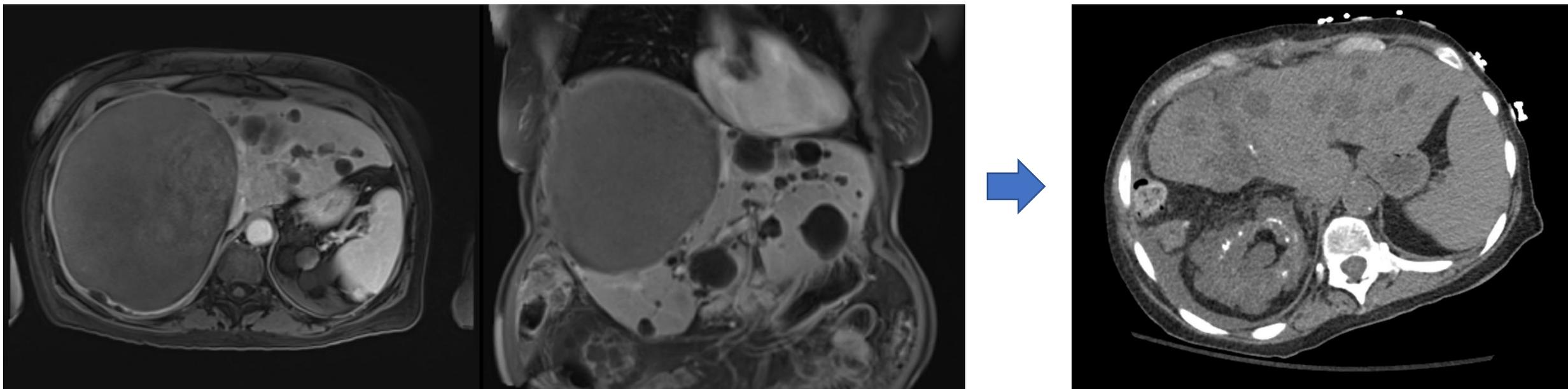
Cyst distribution with relative sparing of right or left lobe

Reported post op morbidity rate 21-51%

Post op adhesions complicate candidacy for liver transplantation

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Polycystic Liver Disease



68 F with longstanding abdominal pain, progressively worse. Severe shortness of breath and pain with activity. Favorable cyst distribution, failed prior percutaneous drainage. Right hepatectomy performed with significant relief of symptoms.

Polycystic Liver Disease

"I want to be cured"

Liver Transplantation

Liver transplantation is only curative treatment option

Only a minority of patients will qualify for transplant

5 year graft/patient survival 87.5% and 92.3% respectively

Complex transplant, preserved liver function (low MELD)

Limited MELD exception points based on weight loss, malnutrition, frailty

If concurrent PKD, consider combined liver-kidney transplant

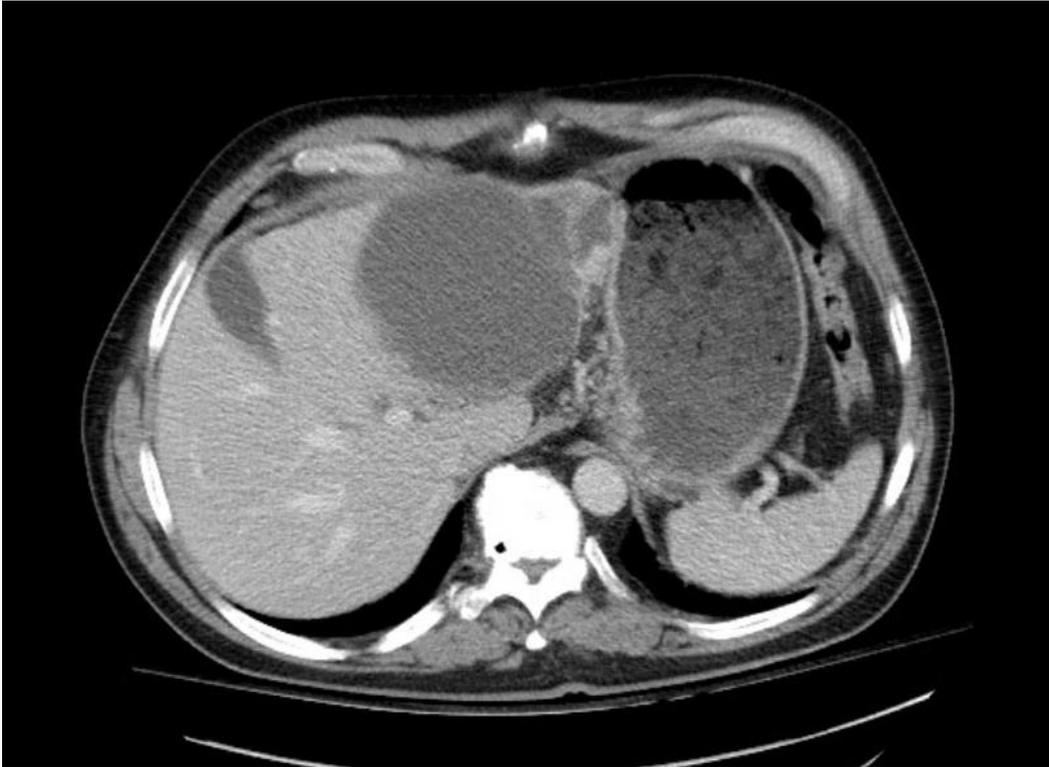
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Polycystic Liver Disease

Listed for Liver Transplantation



Cystic Liver Metastases



72 male with progressive severe epigastric pain. History of prostate cancer.

No extrahepatic disease noted on imaging or screening endoscopy

Left lobe liver resection: **Metastatic GIST tumor**

Octreotide scan negative, no primary identified

Surveillance imaging on imatinib

Cystic Liver Metastases

Important diagnosis to consider, often multiple lesions are present

Common primary sources are colon, kidney, prostate, ovary/testis, squamous cell lung cancer, GIST and NET

50% of all cystic liver metastases arise from colon cancer

Cystic appearance due to development of central necrosis

Liver abscess important differential diagnosis – correlate clinically

Infectious Cysts

Hydatid cyst

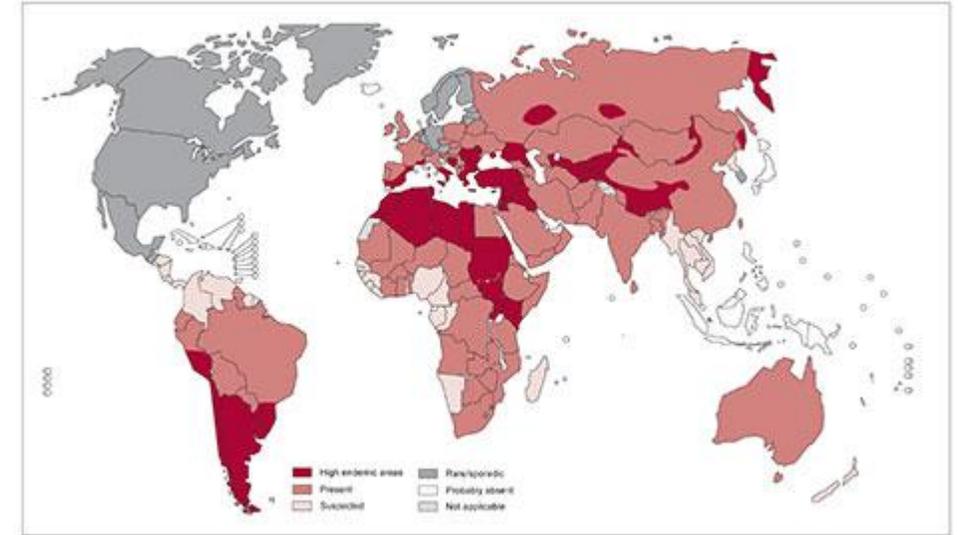
Rare diagnosis in US, identify risk factors based on residence, travel

Often asymptomatic, but may have fever, pruritis, eosinophilia

CT scan can reveal calcified cystic walls, daughter cysts, cyst volume and density

History + imaging features +/- serology
IgE Ab

Distribution of *Echinococcus granulosus* and cystic echinococcosis, worldwide, 2011



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Data Source: World Health Organization
Map Production: Control of Neglected
Tropical Diseases (CNTD)
World Health Organization



Infectious Cysts

Liver Abscess

Unique appearance on CT scan, typically noted by radiology impression

Clinical history important: exclude malignancy, biliary strictures

Antibiotic course and investigation of source, consider diverticulitis

Percutaneous sampling for cultures, cautious about placement of drains without surgical consultation

Hemangioma

Not technically cystic mass, but can be reported as such

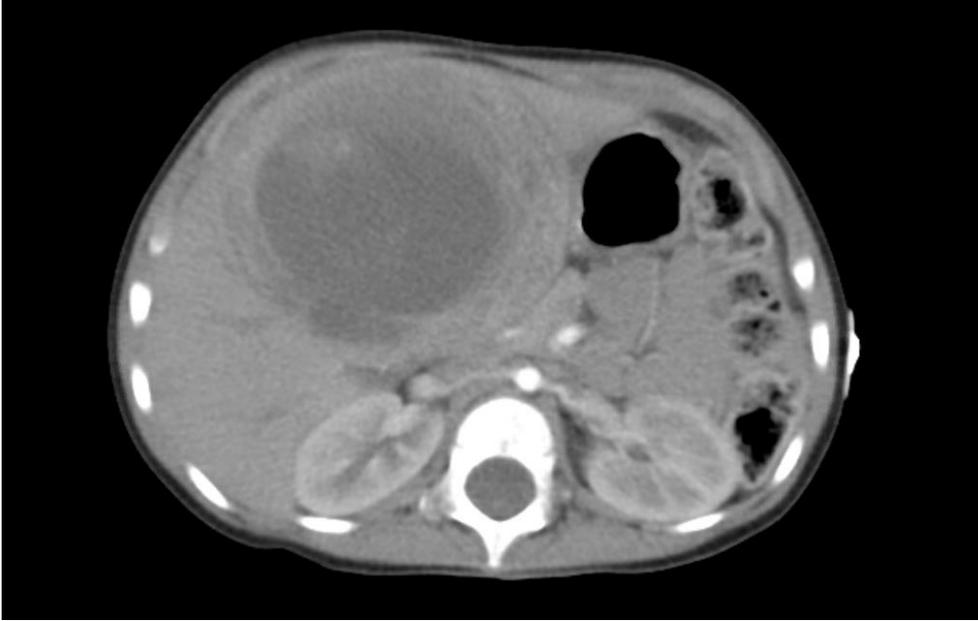
Diagnosis almost always possible from contrast imaging, avoid biopsy

No significant risk of malignant transformation or rupture/bleed

Symptoms primary indication for intervention, again important to carefully assess attribution of pain to hemangioma

Embolization reported as alternative to surgery, but overall low success rates in early experience

Cystic Liver Mass in Kids



Hepatic cysts in children often malignant

Most common embryonal sarcomas

Rare tumors, but should not be sampled/drained

Early referral to pediatric/transplant surgery

2 y/o presented with abdominal fullness. Large cystic mass resected:

Undifferentiated embryonal sarcoma

Adjuvant chemotherapy, doing well

Summary

Small simple cysts: reassurance and no additional studies needed

Cysts with complex features: best managed in multi-disciplinary setting with radiology, advanced endoscopy, surgery

Borderline complexity: surveillance imaging or referral to tertiary care

Symptomatic cysts: carefully assess attribution of pain to cyst, consider surgical referral

Minimally invasive approaches to cyst marsupialization and resection expedite recovery