Cystic Liver Lesions

Liver lesions	Pathophysiology/Histology	Notes		US (the investigation for nonspecific abdominal complaints)	CT	MRI	Follow Up	Progression	Medical Management	When to intervene	Intervention Options		
Simple cyst	- Congenital in nature, arising from aberrant bile ducts that lost communication with biliary tree - Fluid (serous) filled - May develop symptoms with pregnancy - Occur in R > L lobe	- Generally have no septations - Prevalent in 5% of population - 50% are singular - Symptoms develop as a result of: 1. Enlargement (mass effect) 2. Hemorrhage (intracystic bleeding is the MC complication) 3. Infection 4. Rupture		- Anechoic lesion - Well-defined border - No septations - Absence of internal vascularity - A thick or nodular wall raises the suspicion of a cystadenoma but can also represent hemorrhage within the cyst	- Well-circumscribed - Homogenous hypoattenuating - No enhancement with contrast	T1: hypointense T2: hyperintense	- < 4cm + asymptomatic: US Q6-12m - Not necessary if asymptomatic & stable for 2 years - PostOp recurrence < 15%	- May undergo slow growth - Larger cyst have ↑ likelihood of rupture or hemorrhage		- Symptomatic - Rapid growth - Diagnostic uncertainty	Fenestration (open vs lap) or unroofing of the extrahepatic portion	Send cyst wall for histopathology to rule out malignancy	
											Wedge or anatomic resection	Appropriate for: 1. concern of malignancy; 2. multiple cysts when effective	
											Aspiration & injection of Alcohol sclerotherapy	- Appropriate for nonsurgical candidates - Associated with high rates of recurrence	Contraindications: 1. Biliary communication; 2. Peritoneal communication 3. Recent intracyst hemorrhage
Polycystic Liver Disease	- AD transmission - 90% associated with adult PKD - Macro- & microscopically are similar to simple cysts - Prevalence of cysts: - 0% at the age of 20Y - 80% at the age of 60Y	- Gigot classification to determine type (1, 2, or 3) - Need to rule out associated brain aneurysms - LFT are almost always normal - Rare complications include: infection & bleeding		- Gross hepatic enlargement - Multiple cystic lesions in liver ± kidney					- Minimally effective - Avoid estrogen replacement (will cause disease progression) - Somatostatin may ↓ liver volume	Only when symptomatic: - Pain - Infection - Cyst rupture - Hemorrhage - Pressure symptoms - Portal HTN	Open fenestration: appropriate for a few large cysts + ⊖ liver enlargement	Complication rate: 30%	Symptom recurrence: 35%
											Resection ± fenestration: for massive cystic disease	Complication rate: 65%	
											Transplantation	PLD has a MELD Exception criteria	
											Branch embolization may increase parenchymal:cyst ratio	Very time & labor intensive	
											Percutaneous sclerotherapy fails in most		
Ciliated Hepatic Foregut Cysts	Four-layered border with ciliated columnar epithelial lining ± secretes fluid	- Exceedingly rare - Usually single cyst, unilocular - CA19-9 may be ↑		4-Layered wall				Benign, but has malignant potential with larger cysts		1. Pain 2. > 4 cm 3. ⊕ wall abnormalities 4. Enlarge rapidly	Resection		
Traumatic	May be associated with a ruptured liver lesion (adenoma or HCC)			Cystic liver lesions with variable density and intensity; density layering shifts with movement		T1: hyperintense T2: hypointense				If ruptures or complications develop (bile leak, persistent pain, bleeding, compression)			
Hepatic Adenoma	- Benign proliferation of hepatocytes (rich in fat & glycogen) with absence of Kupffer & bile ductules in a background of normal liver - Well-circumscribed - Lack a defined capsule - Necrosis may occur as it outgrows its blood supply - Uncommon (< 30%) to find multiple adenomas. - The main risk factor is use of OCP (↑ with duration & dosage)	- Rare tumors - Occurs in ♀ 20-40Y; rare in ♂ - Occur in R > L lobe - Most are asymptomatic - May develop spontaneous rupture and hemorrhage, possibly fatal - Seen frequently in patients with glycogen storage diseases types I and III - This is the only benign tumor that has definitely been proven to be responsive to OCP	Have 4 subtypes - Beta-catenin mutation is a risk for developing HCC - Inflammatory: have the highest risk of rupture - HNF1α-inactivated have the lowest risk of malignancy - Unclassified Bx is not indicated to identify the subtype	- Hypoechoic or hyperechoic - Can be confused for malignancy	- Well-encapsulated - Nonenhanced: isointense - Enhancement during arterial phase - Peripheral enhancement with centripetal progression - Late phase: may become isodense and then hypodense - Hyperintense with active bleeding	T1: hyperintense T2: hyperintense Shows content of fat or hemorrhage Usually shows well demarcation because of the fat or glycogen content	- AFP if suspecting HCC transformation	- ↑ in size & possible rupture - HCC transformation occurs in 4% of adenomas > 5 cm	If < 5 cm: cessation of OCP & steroids → regress; but this is controversial	- > 5 cm - Symptomatic - Growth off OCP - Patients with complications - β-catenin mutated - Rising AFP - Male patients	Resection (to ↓ risk of malignancy & rupture)	Pringles maneuver helps control bleeding	
											RFA	Appropriate for small or multiple adenomas not amenable to surgery	
										Resection prior to planned pregnancy is usually recommended	Hepatic artery embolization is a temporizing measure (followed by resection during same admission)	Appropriate if actively bleeding + stable patient	
Cystadenoma	- Occur almost exclusively in ♀ - 85% Demonstrate ovarian-like stroma & papillary epithelial projections - Content: serious to mucinous - Content: may show ↑ CA19-9 & CEA - 3 Layer wall: epithelium, mesenchymal stroma, outer CT or pseudocapsule	- The MC primary cystic tumor of the liver - Tend to be large: 10-20 cm - Mainly affect women > 40Y - Lobulated > unilocular - Rarely multiple		- Cystic structure with varying wall thickness, nodularity, septations, and fluid-filled locules	- Hypodense - Multiloculated - Enhancement of cyst wall & septae - Variable fluid attenuation - ± Calcification	More reliable in demonstrating septations/ loculations T1: hyporintense T2: hyperintense		- Grow slowly - High malignant potential (cystadenocarcinoma) - Have great size potential (↑ girth & compressive symptoms)		Always	Partial hepatectomy	IOC may be needed to R/O biliary communication	
											Enucleation		
Cysteadenocarcinoma	- Arise from malignant transformation of a cystadenoma - My occur with or without ovarian-like stroma	- Extremely rare malignancy - Diagnostic aspiration has high risk of peritoneal seeding - Patients develop pain and local compression symptoms - Tumor may rupture - Tend to lack bile duct communication. If present, suspect IPMN-B or cystic cholangiocarcinoma									Hepatic resection or wedge resection preferred over enucleation	- IOC is recommended to R/O biliary communication - Tumor spillage will lead to carcinomatosis	
IPMN-B	- Biliary cystic neoplasms are similar to IPMN i.e (IPMN-B) - Types: - Cystic IPMN-B: large + cystic - Ductectatic IPMN-B - Develop septations & calcificaitons	- May resemble cystadenomas or cystadenocarcinoma - Mostly asymptomatic	ERCP & IOC must be done to demonstrate biliary luminal communication, if present	- Multilocular - Septations - Calcifcations - Nodules - Bile duct dilatation		MRCP will not demonstrate narrow biliary luminal communication		- Extremely high malignant potential - Invasive adenocarcinoma found in 60% of cystic IPMN-B		All cystic IPMN-B	Hepatectomy (preferred)	Done because of its tendency for superficial spread along the bile duct lumen	
											Enucleation	Insufficient & has ↑ recurrence	
Hydatid Disease	- Endemic in Africa, Middle East, South America, Asia, parts of Europe	- 50% are asymptomatic - Symptoms: biliary obstruction (2/2 to rupture or compression); portal HTN, Budd-Chiari (venous outflow obstruction); secondary infection; anaphylaxis - Sr.Antigen ELISA sensitivity 85-90%; cross-reactive for other cestode or helminthic infections			- Excellent characterization of cyst & relation to structures - Great for operative planning	- Preferred to CT in ruling out amebic cysts, congenital cysts, & cystadenoma - MRCP can evaluate cystobiliary communication	- Q3-6 months: - LFTs (albendazole) - CBC (albendazole)		- Albendazole BID - Teratogenic - Hepatotoxic - ↓WBC & ↓Plt - Adding praziquantel Qweek improves killing - < 5 cm uncomplicated (CE1-3) respond well —otherwise, success is 30%				
Cystic Echinococcus	- Is the predominant form - Caused by E. Granulosus - 80% involve 1 organ: 80% liver & 20% lung - Cloudy/discolored fluid suggests communication with biliary tree or secondary infection - WHO Dx: - Possible Dx = clinical history + (imaging or seropositivity) - Probable Dx = clinical + imaging + 2 ⊕ serologies - Confirmed Dx = probable Dx + (microscopic demonstration of protoscolices **or** US findings showing progression of WHO classification)			WHO Classification: - CL = wall not visible - CE1 (active) = Unilocular, anechoic, wall visible - CE2 (active) = Multivesicular - CE3 (transitional) = detached floating membrane, less round - CE4 (inactive) = degenerating membrane, no daughter cyst - CE5 (inactive) = thick calcific wall, arch-shaped			US Q3-6 months to assess recurrence				US-guided PAIR - Done through the liver to avoid intraperitoneal spillage - Agents used: 20% HTS or 95% ethanol - Appropriate for cyst that are: 1. < 5 cm 2. Unilocular (CE1 & CE3a) 3. Accessible percutaneously - WHO recommends starting Rx > 4h before PAIR & continue 1m after - Leave a drain in if >10 cm; DC drain when output < 10 ml/d		- Absolute contraindications: 1. Complicated cyst: infection or rupture 2. Evidence of biliary or pulmonary communication - Morbidity rate 8-13%
											Surgery is the gold standard; - May be open or laparoscopic - Laparoscopic has slightly ↑risk of spillage - Choice: radical > conservative surgery	-Recommended for - Cysts with infection - Communication with biliary tree - Superficial cyst risking rupture - Multiple daughter cysts - Cysts exerting pressure on adjacent organs - Morbidity 30%; mortality up to 4%	- Suspect cyst-biliary fistula if - Fluid is bile stained - Size > 7.5 cm - The communication will need to be ligated with absorbable sutures followed by omentoplasty - ERCP sphincterotomy is therapeutic if bile leak persists postoperatively, ± PTC
											Conservative: partial or subtotal resection of the cyst	- Endocyst is resected - Pericyst is left behind — when packed with omentum, decreases morbidity & recurrence - Use of external drains has now fallen out of fashion	
											Radical: - Pericystectomy - Segmental lobe resection - Rarely, transplantation	- Remove entire cyst	2% develop recurrence
Alveolar Echinococcus	- Caused by E. Alveolar - High affinity to the liver - Behave tumor-like with invasive noncystic growth	- Aggressive infiltrative disease: - Biliary obstruction - Budd-Chiari - Liver failure - Has PNM staging - P: extension of parasitic disease - N: neighboring organ involvement - M: extrahepatic involvement		- Pseudotumor - ± hemangioma-like appearance - ± small calcifications			CT or MRI Q1-2Y		Recommended for at least 2 years, possibly indefinitely if inoperable		- Percutaneous management is not an option - Surgery is mandated - Goal: R0 resection - No role for debulking or paliative procedures		
Hemangioma	- Unknown pathogenesis: congenital vs acquired - Hormonal association has been suggested - Have a layer of endothelium lining surrounded by thin CT ± calcification & fibrosis - Blood supply is derived from the hepatic artery - Have no metastatic potential - Very low risk of spontaneous rupture or hemorrhage - Large hemangiomata in children can result in CHF secondary to AV shunting. Untreated symptomatic childhood hemangiomata are associated with a 70% mortality	- It is the MC benign tumor of the liver - ♀ 4:1 ♂ - ⊕ in 20% of population - Most are asymptomatic - 50% have multifocal lesions - Usually < 5 cm - Symptoms: - Pain: thrombosis vs stretch of Glisson's capsule - Mass effect: jaundice, GOO - AV shunting → CHF - Life Threatening: - Rupture - Hemorrhage - Kasabach-Merritt syndrome: hemangioma traps Plt, leads to DIC. The syndrome carries 30% mortality		- Well-defined - Hyperechoic homogeneous mass - ± Central necrosis & fibrosis	- Multiphase CT - Arterial: Peripheral nodular enhancement (periphery) - Venous: centripetal filling - Delayed: retained contrast - Above features sufficient for Dx - If regressed or thrombosed, may mimic malignancy	T1: hypointense T2: hyperintense - Usefulness: - When CT is not diagnostic - ↓Radiation on long-term surveillance	- 15% regress - No appropriate algorithm for asymptomatic patients - Acceptable algorithm: MRI 3m after Dx - Repeat in 3-6m if lesion is atypical - ↑CA19-9, CEA & alpha-FP are unusual and warrant further evaluation	↑Rate of growth while on steroids, OCP, or pregnancy - Rapid ↑ Size is unusual: suspect malignancy	Historically, females were advised to avoid pregnancy for fear or rupture; however, this risk is small and evasion of pregnancy is not indicated	- Extreme pain - Mass-effect related symptoms - Intraperitoneal hemorrhage - Inability to R/O malignancy - Kasabach-Merritt Syndrome - Size alone is not an indication for resection - Observation vs operative intervention resulted in a similar incidence of hemangioma related complications	IR embolization (followed by interval resection)	Appropriate 1st line for: 1. Kasabach-Merritt syndrome; 2. Intraperitoneal hemorrhage	
											Enucleation (preferred when possible)	- Associated with ↓blood loss, ↓OR time, ↓complications - Recurrence is rare	Pringle Maneuver is useful; inflow occlusion < 30m is well tolerated, otherwise intermittently unclamp for 5 mins
											Liver resection (anatomic or nonanatomic), (lap or open)	Done when there is rupture or diagnostic uncertainty	Standard liver resection techniques & principles apply
											- Other options: transarterial emoblization; percutaneous RFA, percutaneous ethanol ablation, hepatic radiation therapy - Percutaneous Bx is contraindicated		
Focal Nodular Hyperplasia	- Polyclonal proliferation of normal parenchyma formed as a hyperplastic response to a fibrous scar of vessels found in the center of these lesions - Central scar often contains a large artery branching out into multiple smaller arteries in a spoke wheel pattern - 20% are atypical: have no central scar - OCP is associated with larger, more symptomatic, & vascular lesions	- The 2nd MC benign hepatic lesion - Usually a single lesion < 5 cm - Most common in ♀ - Have no potential for - Malignancy - Rupture or hemorrhage - When central scar is not seen, it's difficult to differentiate from fibrolamellar HCC on imaging - On sulfur colloid scan, it's the only lesion that is not "cold"	Percutanous Bx is not advisable (risks seeding if malignant, and has low accuracy for Dx), but is done sometimes in diagnostic uncertainties	- Hyperechoic, isoechoic, or hypoechoic - Central scar is hard to see on US	- Well-circumscribed - Arterial: rapid enhancement - Venous: isointense + hyperintense central scar	T1: hypointense T2: hyperintense	Asymptomatic: short-term surveillance		Stopping OCP is controversial	- Pain - Weight loss - Inability to R/O malignancy (often in atypical FNH)	Resection		
Other	**Secondary Cystic Neoplasms:** Seen with metastasis of: 1. Bronchus & breast carcinoma, 2. Ovarian & pancreatic cystadenocarcinoma				**Embryonal Hepatic Sarcoma**: Occur at age 2-15Y; have both myxoid and necrotic contents				**HCC & cholangiocarcinoma** may experience cystic changes				