Manchester Cancer

Central Manchester University Hospitals NHS
NHS Foundation Trust

Greater Manchester and Cheshire HPB Unit Guidelines for the Assessment & Management of Hepatobiliary and Pancreatic Disease

Chapter 7

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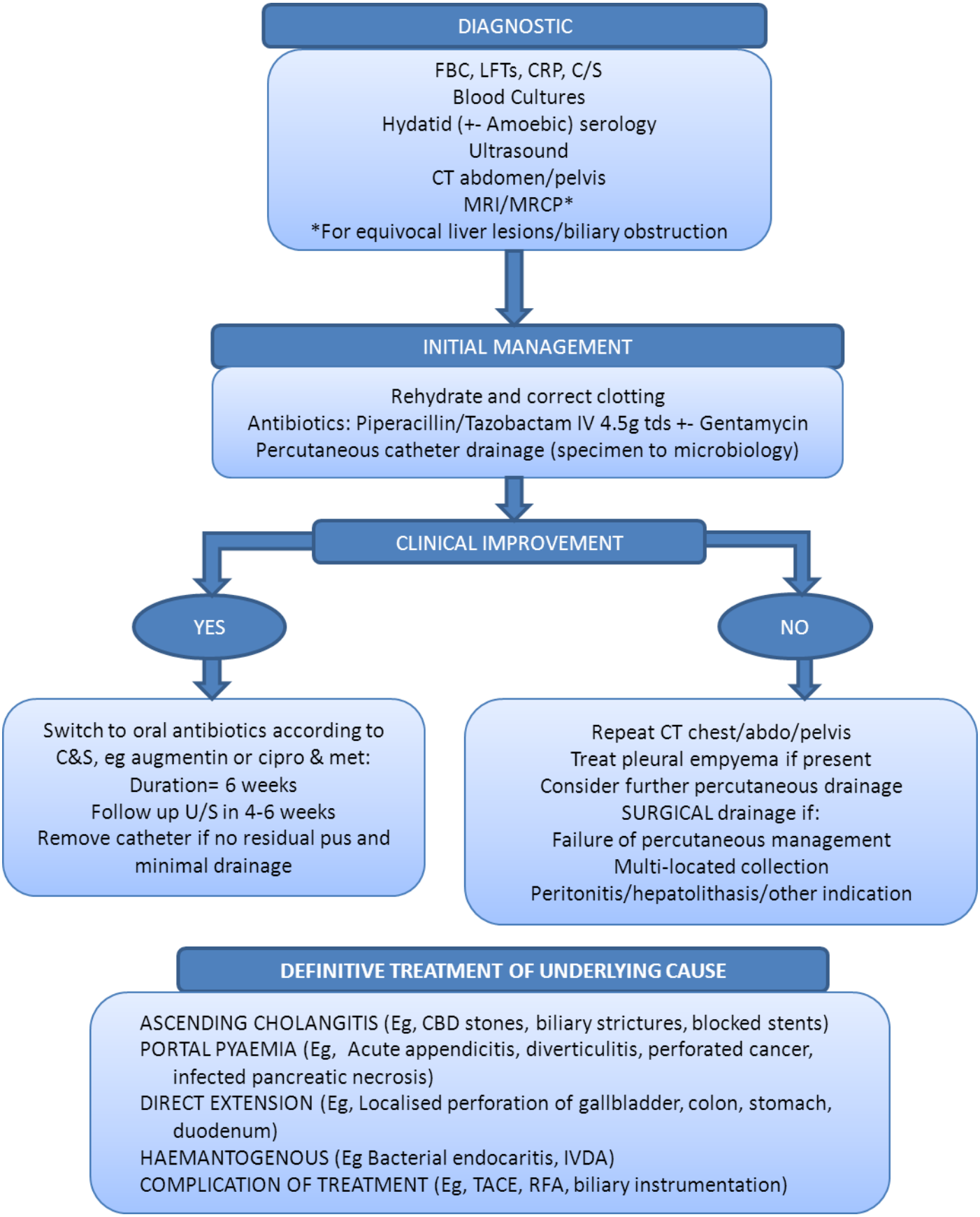
7. Benign liver conditions

7.1. *Pyogenic liver abscess*



7.2. *Management of hydatid cysts (cystic echinococcosis)*

Fig. 1. Diagrammatic representation of structure of the echinococcal cyst.

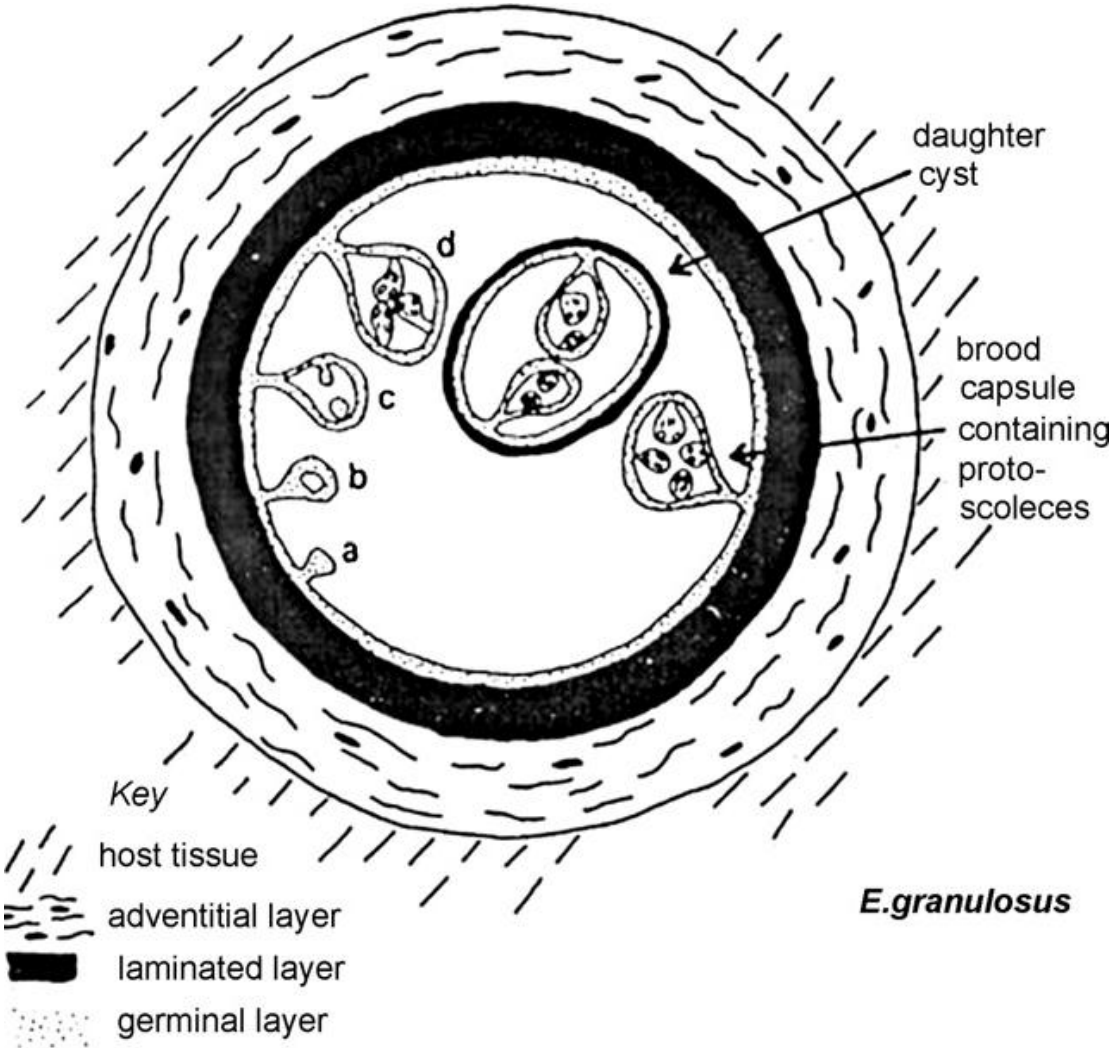
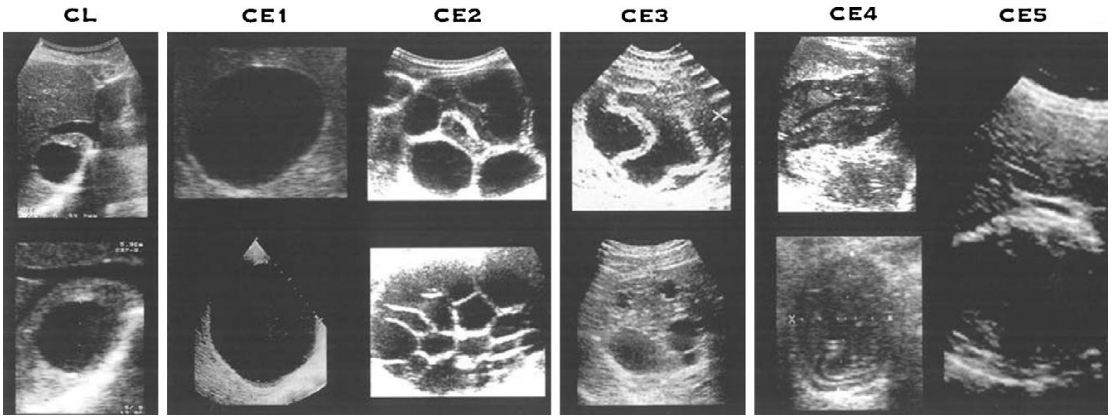


Fig. 2. WHO-IWGE CLASSIFICATION OF CYSTIC ECHINOCOCCOSIS



The WHO-IWGE standardised classification allows a natural grouping of the cysts into three relevant groups: active (CE1 and 2), transitional (CE3) and inactive (CE4 and 5). It includes a “cystic lesion” (CL) stage (undifferentiated).

Imaging Features of Hydatid Cysts:

Cystic Lesion

- Status: if cystic echinococcosis [CE] active
- Unilocular cystic lesion [CL] with uniform anechoic content, not clearly delimited by hyper echoic rim (= cyst wall not visible)
- Normally round but may be oval
- Size variable often small; CLs (< 5.0cm), but may be medium sized (CLm (5-10cm), or large (CLl >10cm)
- Normally these are non-parasitic cystic lesions, but if there is a suspicion of CE, these cysts are usually at an early stage of development and are not fertile. US does not detect any pathognomonic signs. Differential diagnosis of these cystic lesions requires the application of the additional diagnostic techniques.

Cystic Echinococcosis

- Active
- Unilocular simple cyst with uniform anechoic content. Cyst may exhibit fine echoes due to shifting of brood capsules which is often called hydatid sand or “snowflake sign”
- Cyst wall is visible
- Normally round or oval
- Size variable: CE1s (<5.0cms), CE1m (5-10cms), CE1l (>10cm)
- Usually fertile – pathognomonic signs include visible cyst wall and snowflake sign

CE2

- Status: Active
- Multivesicular, multiseptated cysts in which the daughter cysts may partly or completely fill with unilocular mother cysts. Cyst septations may produce “wheel-like” structures or the contained daughter cysts may produce a “rosette-like” or “honeycomb” structure.
- Cyst wall normally visible
- Normally round or oval
- Size variable: CE2s (<5.0cms), CE2m (5-10cms), CE2l (>10cm)
- Usually fertile
- US features are pathognomonic

CE3

- Status: Transitional
- Anechoic content with detachment of laminated membrane from, the cyst wall visible as floating membrane or as “water-lily sign”: which is indicative of wavy membranes floating on top of the remaining cyst fluid
- Unilocular cyst which may contain daughter cysts (anechoic appearance) and echoic areas (disrupted membranes/degenerating daughter cysts). These cysts appear at US as a “complex mass”
- Cyst form may be less rounded due to decrease of intra-cystic fluid pressure
- Size variable: CE3s (<5.0cms), CE3m (5-10cms), CE3l (>10cm)
- Transitional stage: Cyst is most usually starting to degenerate. Degenerative signs of US examination are “detachment and rupture of membranes”. Occasionally may be followed by daughter cyst production
- US features are pathognomonic

CE4

- Status: Inactive
- Heterogenous hyperechoic or dyshomogenous degenerative contents. No daughter cysts
- May show a “ball of wool” sign which is indicative of degenerating membranes
- Size variable: CE4s (<5.0cms), CE4m (5-10cms), CE4l (>10cm)
- Most cysts of this type are not fertile
- US features are usually not pathognomonic and further diagnostic tests are required to confirm the diagnosis. Differential diagnosis may be possible if there is presence of a cystic wall, lateral cone shadow, little calcifications, or if an echoic and anechoic spiral “ball of wool” image is clearly seen within a focal hepatic lesion

CE5

- Status: Inactive
- Cysts are characterised by thick walled calcified wall which is arch shaped producing a cone shaped shadow. Degree of calcification varies from partial to complete.
- Size variable: CE5s (<5.0cms), CE5m (5-10cms), CE5l (>10cm)
- Cysts not fertile in majority of cases
- Diagnosis uncertain, features are not pathognomonic, but highly suggestive of E. granulosus.

Figure 3. Diagnostic algorithm for hydatid liver cysts

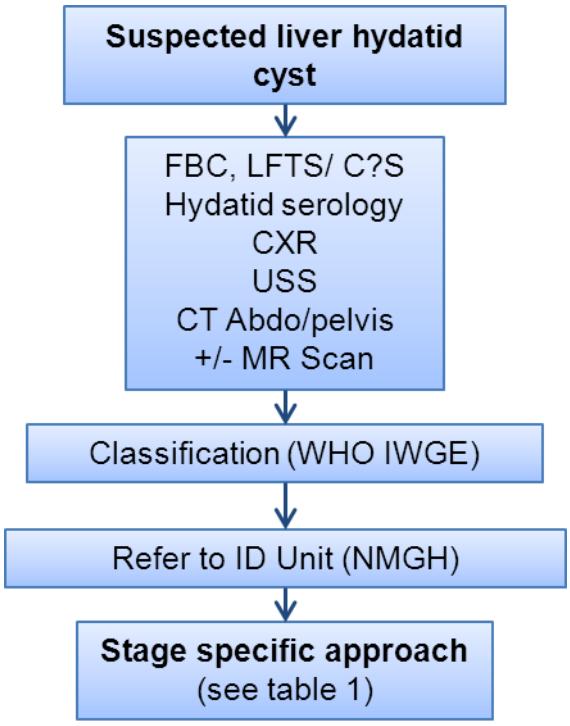
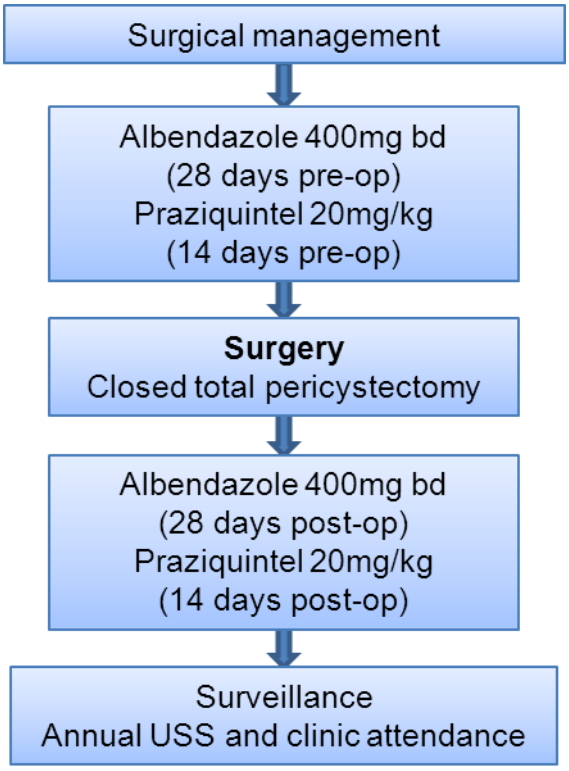


Table 1. WHO-IWGE suggested stage-specific approach to uncomplicated cystic echinococcosis of the liver

WHO classification	Surgery	Percutaneous treatment	Drug therapy	Suggested	Resources
CE1				<5cm ABZ	Optimal
				PAIR	Minimal
		√	√	>5cm PAIR + ABZ	Optimal
				PAIR	Minimal
CE2	√			Other PT + ABZ	Optimal
		√	√		
				Other PT	Minimal
CE3a				<5cm ABZ	Optimal
		√	√	PAIR	Minimal
				>5cm PAIR + ABZ	Optimal
				PAIR	Minimal
CE3b				Non-PAIR PT + ABZ	Optimal
	√	√	√		
				Non-PAIR PT	Minimal
CE4				Watch and Wait	Optimal
CE5				Watch and Wait	Optimal

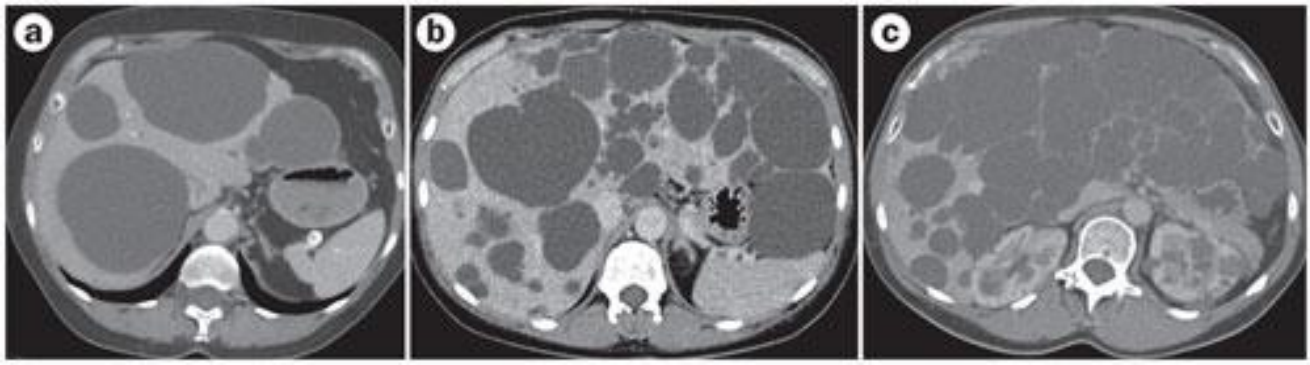
PAIR; Puncture, aspiration, injection, re-aspiration.

Figure 4. Surgical management of active and transitional hydatid cysts



7.3. *Solitary & Polycystic liver disease*

Transverse CT images of polycystic liver disease

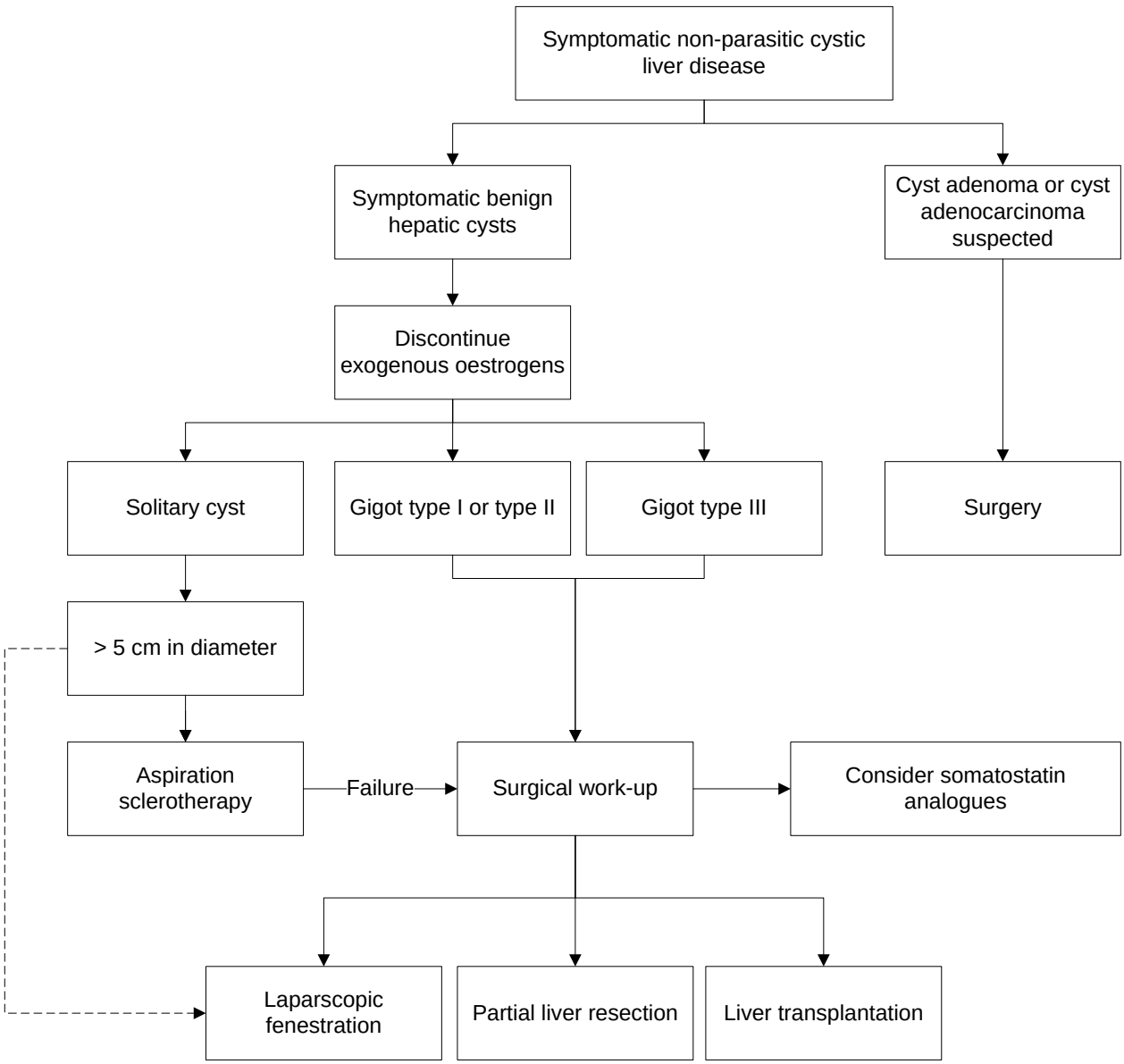


a | Gigot type I cystic liver containing a couple of large (>10 cm) cysts, but <10 cysts in total.

b | Gigot type II polycystic liver with diffuse involvement of liver parenchyma by multiple medium-sized cysts.

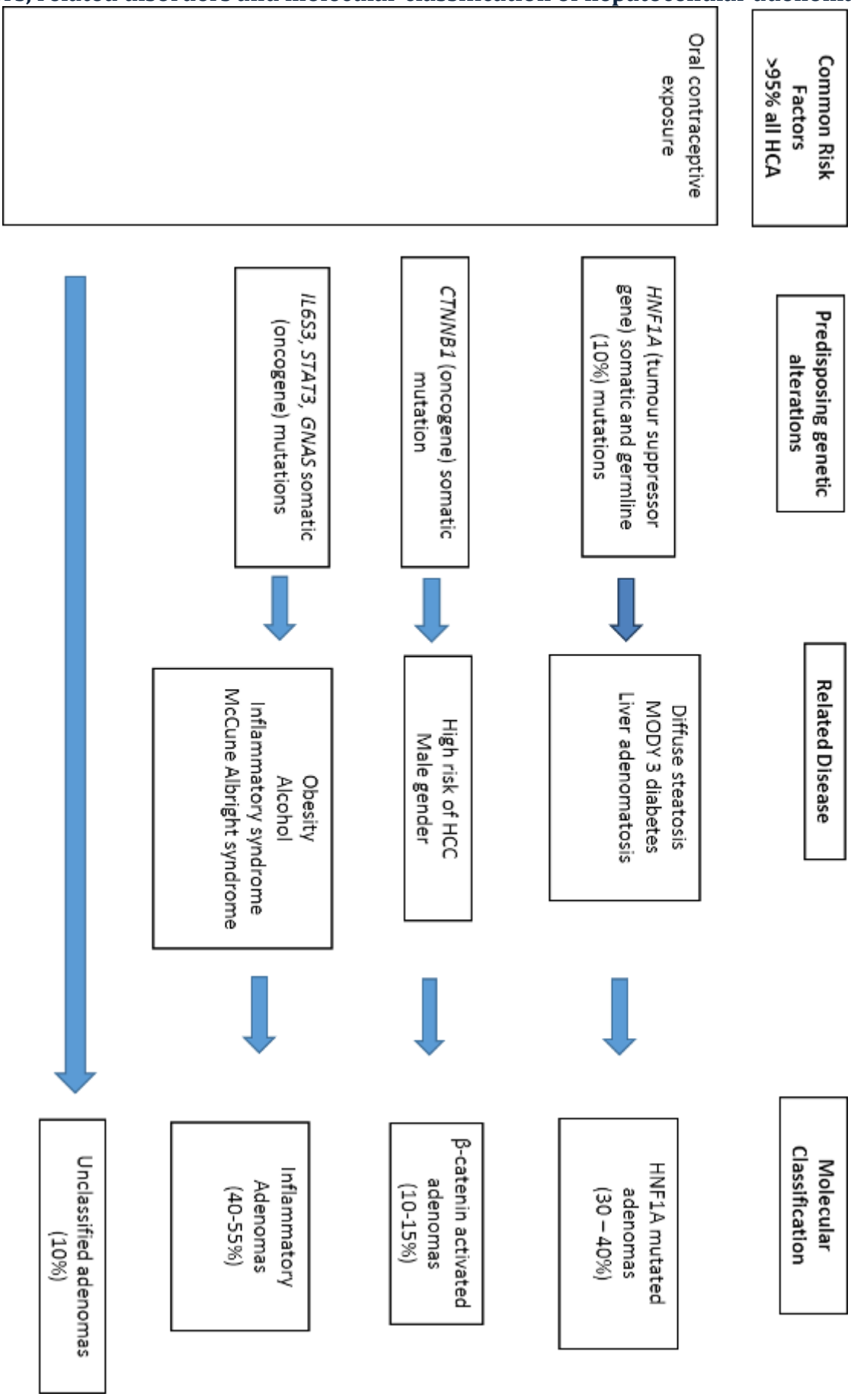
c | Gigot type III polycystic liver. The liver is completely occupied with numerous cysts, and only a few areas of visible liver parenchyma are present.

Management pathway for symptomatic non-parasitic benign cystic liver disease

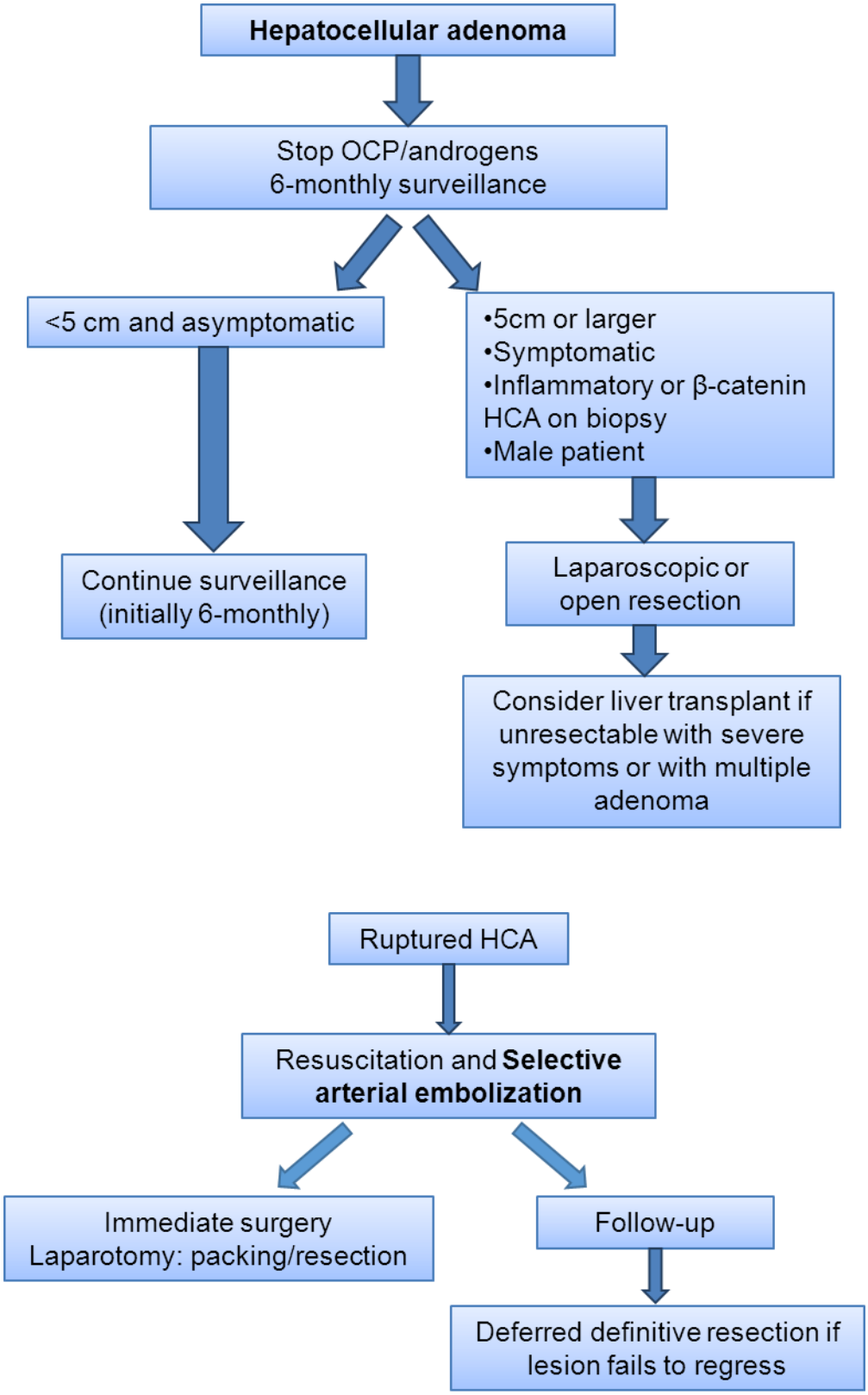


7.4. *Hepatocellular adenomas*

Risk factors, related disorders and molecular classification of hepatocellular adenoma



Management algorithms for stable and ruptured hepatocellular adenoma



7.5. *Acute Liver Failure*

Common Management Issues

Organ system and Common Conditions	Assessment	Specific Elements of Care
Cardiovascular system		
Hypotension	Invasive monitoring for all conditions; echocardiography for low cardiac output and right ventricular failure	
Intravascular volume depletion		Correction of volume depletion
Vasodilation		Vasopressors
Low cardiac output and right ventricular failure		Inotropic support
Hepatic system		
Evolving hepatic dysfunction	Serial biochemical and coagulation testing	Intravenous acetylcysteine
Respiratory system		
Risk of aspiration pneumonia	Neurologic observation to monitor level of consciousness	Early tracheal intubation for depressed level of consciousness
Metabolic and renal systems		
Hypoglycaemia	Serial biochemical testing	Maintain normoglycaemia
Hyponatraemia		Active fluid management
Renal dysfunction, lactic acidosis, hyperammonaemia		Renal-replacement therapy
Impaired drug metabolism		Review drug administration
Central nervous system		
Progressive encephalopathy	Neurologic observation; monitoring of serum ammonia level; transcranial ultrasonography; consideration of intra-cranial pressure monitoring	Treatment of fever and hyponatraemia; screening for sepsis High-grade encephalopathy; endotracheal intubation; avoidance of PaCO2 od <30mmHG or >45 mmHg; target for serum sodium, 145-150 mmol/l; risk assessment for intra-cranial hypertension
Intracranial hypertension		Interventions for pressure surges; osmotherapy (mannitol, hypertonic saline); temperature control; rescue therapies (indomethacin, thiopentone)
Haematologic system		
Coagulopathy	Laboratory coagulation testing	No routine correction of coagulation abnormalities, only for invasive procedures
Immunologic system		
High risk of sepsis	Clinical evaluation	Antibiotic prophylaxis

West Haven criteria for grading mental state in hepatic encephalopathy

Grade	Features
Grade 0	No signs or symptoms
Grade 1	Euphoria, anxiety, trivial lack of awareness, impaired performance, shortened attention span, mild asterixis
Grade 2	Lethargy, minimal personality changes, subtle personality change, inappropriate behaviour, asterixis
Grade 3	Somnolence, confusion, gross disorientation
Grade 4	Coma

King's College criteria for liver transplantation in Acute Hepatic Failure.

Acetaminophen-associated AHF	All other causes of AHF
pH < 7.3	INR >6.5
Or	Or
INR > 6.5, serum creatinine >3.4 mg/dl, and grade III–IV encephalopathy	Three of the following variables:
	1. Age <10 or >40 years
	2. Cause is non-A, non-B hepatitis or idiosyncratic drug reaction
	3. Duration of jaundice before encephalopathy >7 days
	4. INR > 3.5
	5. Serum bilirubin >17.5 mg/dl

7.6. *Liver Transplantation: UK Selection Criteria*

Liver Advisory Group on behalf of NHSBT

http://www.odt.nhs.uk/pdf/liver_selection_policy.pdf

Conditions that are considered for transplantation

Adult patients

Most adult patients with liver disease are not managed in transplant centres. Patients referred for assessment for liver transplant will include those with the following broad categories of conditions:

Acute liver failure

- Multi-system disorder in which severe acute impairment of liver function with encephalopathy occurs within 8 weeks of the onset of symptoms and no recognised underlying chronic liver disease

Chronic liver disease; any cirrhosis which may be due to:

- Alcoholic liver disease
- Non-alcoholic fatty liver disease
- Chronic viral hepatitis B, C or D
- Autoimmune liver diseases: primary biliary cirrhosis, primary sclerosing cholangitis, chronic active liver disease and overlap syndromes
- Genetic haemochromatosis
- Wilson's disease
- Alpha-1 antitrypsin deficiency
- Congenital hepatic fibrosis and other congenital or hereditary liver diseases
- Secondary sclerosing cholangitis

Liver tumours

- Hepatocellular carcinoma (See: *6.7 Hepatocellular Cancer - Liver Transplantation*)

Variant syndromes

- Diuretic resistant ascites
- Chronic hepatic encephalopathy
- Intractable pruritus
- Hepatopulmonary syndrome
- Familial amyloid polyneuropathy
- Familial hypercholesterolaemia
- Polycystic liver disease
- Hepatic epithelioid haemangioendothelioma
- Sickle cell hepatopathy

Patients not falling within these categories may be considered through the National Appeals Panel route.