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7. Benign liver conditions
7.1. *Pyogenic liver abscess*

**DIAGNOSTIC**
- FBC, LFTs, CRP, C/S
- Blood Cultures
- Hydatid (+- Amoebic) serology
- Ultrasound
- CT abdomen/pelvis
- MRI/MRCP*
  *For equivocal liver lesions/biliary obstruction*

**INITIAL MANAGEMENT**
- Rehydrate and correct clotting
- Antibiotics: Piperacillin/Tazobactam IV 4.5g tds +- Gentamycin
- Percutaneous catheter drainage (specimen to microbiology)

**CLINICAL IMPROVEMENT**
- **YES**
  - Switch to oral antibiotics according to C&S, eg augmentin or cipro & met:
    - Duration: 6 weeks
    - Follow up U/S in 4-6 weeks
    - Remove catheter if no residual pus and minimal drainage
- **NO**
  - Repeat CT chest/abdo/pelvis
  - Treat pleural empyema if present
  - Consider further percutaneous drainage
  - SURGICAL drainage if:
    - Failure of percutaneous management
    - Multi-located collection
    - Peritonitis/hepatolithiasis/other indication

**DEFINITIVE TREATMENT OF UNDERLYING CAUSE**
- **ASCENDING CHOLANGITIS** (Eg, CBD stones, biliary strictures, blocked stents)
- **PORTAL PYAEMIA** (Eg, Acute appendicitis, diverticulitis, perforated cancer, infected pancreatic necrosis)
- **DIRECT EXTENSION** (Eg, Localised perforation of gallbladder, colon, stomach, duodenum)
- **HAEMANTOGENOUS** (Eg Bacterial endocarditis, IVDA)
- **COMPLICATION OF TREATMENT** (Eg, TACE, RFA, biliary instrumentation)
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

<table>
<thead>
<tr>
<th>WHO classification</th>
<th>Surgery</th>
<th>Percutaneous treatment</th>
<th>Drug therapy</th>
<th>Suggested</th>
<th>Resources</th>
</tr>
</thead>
<tbody>
<tr>
<td>CE1</td>
<td></td>
<td></td>
<td>&lt;5cm ABZ</td>
<td></td>
<td>Optimal</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>PAIR</td>
<td></td>
<td>Minimal</td>
</tr>
<tr>
<td></td>
<td>✓</td>
<td>✓</td>
<td>&gt;5cm PAIR + ABZ</td>
<td>Optimal</td>
<td>Minimal</td>
</tr>
<tr>
<td>CE2</td>
<td>✓</td>
<td>✓</td>
<td>Other PT + ABZ</td>
<td>Optimal</td>
<td>Minimal</td>
</tr>
<tr>
<td></td>
<td>✓ ✓</td>
<td>✓</td>
<td>✓</td>
<td></td>
<td></td>
</tr>
<tr>
<td>CE3a</td>
<td></td>
<td></td>
<td>&lt;5cm ABZ</td>
<td></td>
<td>Optimal</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>PAIR</td>
<td></td>
<td>Minimal</td>
</tr>
<tr>
<td></td>
<td>✓ ✓</td>
<td>✓</td>
<td>&gt;5cm PAIR + ABZ</td>
<td>Optimal</td>
<td>Minimal</td>
</tr>
<tr>
<td>CE3b</td>
<td></td>
<td></td>
<td>Non-PAIR PT + ABZ</td>
<td>Optimal</td>
<td></td>
</tr>
<tr>
<td></td>
<td>✓ ✓</td>
<td>✓</td>
<td>✓</td>
<td></td>
<td></td>
</tr>
<tr>
<td>CE4</td>
<td></td>
<td></td>
<td>Watch and Wait</td>
<td></td>
<td></td>
</tr>
<tr>
<td>CE5</td>
<td></td>
<td></td>
<td>Watch and Wait</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

PAIR; Puncture, aspiration, injection, re-aspiration.
Figure 4. Surgical management of active and transitional hydatid cysts

- Surgical management
  - Albendazole 400mg bd (28 days pre-op)
  - Praziquantel 20mg/kg (14 days pre-op)

- Surgery
  - Closed total pericystectomy

- Albendazole 400mg bd (28 days post-op)
- Praziquantel 20mg/kg (14 days post-op)

- Surveillance
  - Annual USS and clinic attendance
7.3. **Solitary & Polycystic liver disease**

Transverse CT images of polycystic liver disease

![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.

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Management pathway for symptomatic non-parasitic benign cystic liver disease

Symptomatic non-parasitic cystic liver disease

Symptomatic benign hepatic cysts

Discontinue exogenous oestrogens

Solitary cyst

Gigot type I or type II

Gigot type III

Cyst adenoma or cyst adenocarcinoma suspected

Surgery

Aspiration sclerotherapy

Laparoscopic fenestration

Partial liver resection

Liver transplantation

Discontinue exogenous oestrogens

Failure

Surgical work-up

Consider somatostatin analogues

Liver transplantation
7.4. Hepatocellular adenomas

Risk factors, related disorders and molecular classification of hepatocellular adenoma

- Classification
  - Molecular
  - Related Disease

- Factors
  - Common Risk
  - >95% all HCA
  - >40% metalled adenomas
  - (40-50%)
  - Inflammatory adenomas

- Related Adenomas
  - (10-15%)
  - p-catenin activated

- Unrelated Adenomas
  - (10%)
  - HNF4A mutated

- Molecular
  - CMMNN1 (oncogene, somatic)
  - Tumor suppressor
  - (10%) mutations
  - Gene 3 (ATM) and Germline
  - HNF4A (tumor suppressor)

- Inflammatory Adenomas
  - Alcohol
  - Obesity
  - Inflammatory syndrome

- Risk factors
  - IL6ST, ST4473, EN155 somatic

- Adenoma
  - High risk of HCC
  - Male gender

- Classification
  - Molecular
  - Related Disease

- Factors
  - Common Risk
  - >95% all HCA
  - >40% metalled adenomas
  - (40-50%)
  - Inflammatory adenomas

- Related Adenomas
  - (10-15%)
  - p-catenin activated

- Unrelated Adenomas
  - (10%)
  - HNF4A mutated

- Molecular
  - CMMNN1 (oncogene, somatic)
  - Tumor suppressor
  - (10%) mutations
  - Gene 3 (ATM) and Germline
  - HNF4A (tumor suppressor)

- Inflammatory Adenomas
  - Alcohol
  - Obesity
  - Inflammatory syndrome
Management algorithms for stable and ruptured hepatocellular adenoma

1. **Hepatocellular adenoma**
   - Stop OCP/androgens
   - 6-monthly surveillance

   - **<5 cm and asymptomatic**
     - Continue surveillance (initially 6-monthly)

   - **≥5 cm or larger**
     - Symptomatic
     - Inflammatory or β-catenin
     - HCA on biopsy
     - Male patient
     - Laparoscopic or open resection
     - Consider liver transplant if unresectable with severe symptoms or with multiple adenoma

2. **Ruptured HCA**
   - Resuscitation and **Selective arterial embolization**
     - Immediate surgery
       - Laparotomy: packing/resection
     - Follow-up
       - Deferred definitive resection if lesion fails to regress
### 7.5. Acute Liver Failure

**Common Management Issues**

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

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

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

<table>
<thead>
<tr>
<th>Acetaminophen-associated AHF</th>
<th>All other causes of AHF</th>
</tr>
</thead>
<tbody>
<tr>
<td>pH &lt; 7.3</td>
<td>INR &gt;6.5</td>
</tr>
<tr>
<td>Or</td>
<td>Or</td>
</tr>
<tr>
<td>INR &gt; 6.5, serum creatinine &gt;3.4 mg/dl, and grade III–IV encephalopathy</td>
<td>Three of the following variables:</td>
</tr>
</tbody>
</table>

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


**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.