Hepatomegaly

Essential History

Note that the liver is normally palpable 1–3 cm below the costal margin in infants and toddlers. In older children the liver is not normally palpable more than 1 cm below the costal margin.

A palpable liver could either be due to enlargement of the liver or downward displacement of a normal sized liver.

Keywords / also known as: enlarged liver, liver enlargement

Ask about:

- Symptoms or signs suggestive of liver disease (*or haematological disorder)
  - Jaundice*
  - Pale stools
  - Itching
  - Bruising*
  - Petechiae*
  - Epistaxis
  - Haematemesis / melaena
  - Reduced appetite / weight loss
  - Obesity
  - Lethargy*
  - Fever*
  - Thrombotic events*
- Symptoms suggesting metabolic or storage disorder
  - Dietary avoidances
  - Hypoglycaemia
- Symptoms suggesting cardiorespiratory disease
  - Dyspnoea
  - Cough
  - Wheeze
  - Chest pain
- Symptoms suggesting systemic disorder
  - Fever
  - Weight loss
  - Joint symptoms
  - Occular symptoms

  - Rash

Additional history to be considered:

- Perinatal history
  - Including neonatal jaundice, prematurity, umbilical vein cannulation
- Developmental progress
- Family history
- Travel history
- Drug history
- Trauma

'Red Flag' Symptoms and Signs

Ask about:

- Bruising / petechiae
  - Suggests portal hypertension due to liver disease
  - Suggests haematological disorder
- Dyspnoea
  - Suggests cardiac or respiratory disorder
  - Hepatopulmonary syndrome
- Neuropsychiatric symptoms
  - Altered level of consciousness
    - Hepatic encephalopathy
  - Delayed development, loss of skills, altered behaviour
    - May suggest metabolic disorder or Wilson's disease

Look for:

- Splenomegaly
  - Suggests portal hypertension due to liver disease **or** haematological disorder **or** acute viral infection
- Jaundice
  - Suggests liver disease or haematological disorder
  - With pale stools: suggests biliary obstruction (urgent consideration of biliary atresia in infant) or infection such as hepatitis
- Evidence of poor growth (see Faltering Growth)
  - May indicate long term condition

Differential Diagnosis / Conditions

- Biliary obstruction (including biliary atresia in infancy)
- Infective
  - Viral
  - Bacterial
- Toxic (eg, drugs, including paracetamol)
- Autoimmune hepatitis
- Infiltration
- Genetic / metabolic
  - Congenital hepatic fibrosis
  - Cystic fibrosis
  - Wilson's disease
  - Alpha-1-antitrypsin deficiency
  - Alagille's syndrome
  - Storage
    - Fat (non-alcoholic fatty liver disease (NAFLD))
    - Glycogen storage and Mauriac's syndrome
    - Mucopolysaccharidoses
    - Lipid storage disorders
- Cardiorespiratory disorder with downward displacement of liver without true hepatomegaly
  - Hyperinflated lung
    - Asthma
    - Bronchiolitis
    - Pneumonitis
  - Tension pneumothorax
  - Congenital diaphragmatic hernia
  - Thoracic tumours
  - Subdiaphragmatic lesions (eg, abscess)
- Gastro-intestinal conditions
  - Coeliac disease
  - Inflammatory bowel disease
- Malignant tumours
  - Primary tumours
    - Hepatoblastoma
    - Hepatocellular carcinoma
  - Secondary tumours
    - Neuroblastoma
- Benign tumours
  - Focal nodular hyperplasia

  - Mesenchymal hamartoma
  - Haemangioendothelioma
- Haematological malignancies / infiltration
  - Lymphoma
  - Leukaemia
  - Histiocytosis
- Vascular congestion
  - Suprahepatic
    - Congestive heart failure
    - Cardiac tamponade
    - Constrictive pericarditis
  - Intrahepatic
    - Hepatic vein thrombosis (Budd–Chiari syndrome)
    - Hepatic vein web
    - Vascular malformations (cavernous hemangioma, capillary hemangioma, haemangioendothelioma)
  - Sickle cell disease

Investigations

Hepatomegaly especially when accompanied by jaundice, is potentially serious, so investigation and treatment should be discussed with the on-call Paediatrician.

To be undertaken by non-specialist practitioners (eg, General Practitioner (GP) Team) or specialist practitioners (eg, Emergency Department / General Paediatric / Paediatric Gastroenterology Team(s)):

- Depending on clinical assessment, age of child, and associated history and symptoms:
  - Liver enzymes
    - γ-glutamyltransferase
    - Alkaline phosphatase
    - Alanine aminotransferase
    - Aspartate aminotransferase
  - Liver function studies
    - Prothrombin time
    - International normalised ratio
    - Partial thromboplastin time
    - Albumin
    - Total protein
  - Fractionated bilirubin (total and conjugated) and hepatitis screen
  - Blood glucose

  - Full blood count, peripheral blood smear
  - Urine culture
  - Stool culture
  - C-reactive protein / erythrocyte sedimentation rate
  - Ultrasound assessment of liver, spleen, and biliary tree
  - Chest X-ray

To be undertaken by specialist practitioners (eg, Emergency Department / General Paediatric / Paediatric Gastroenterology Team(s)):

- Where appropriate:
  - Hepatitis A, B, C and E serology
  - Cytomegalovirus serology
  - Epstein–Barr virus serology
  - Autoantibodies (LKM, ANA, SMA), and immunoglobulins
  - IgA tissue transglutaminase (tTGA)
    - Coeliac disease
  - Caeruloplasmin
  - Alpha-1-antitrypsin level
  - Lactate dehydrogenase (LDH), Direct antiglobulin test, haptoglobins
    - Haemolysis

Treatment Approach

To be undertaken by non-specialist practitioners (eg, GP Team):

- Treatment is guided by accurate diagnosis, and further diagnostic pathway should be discussed with a Paediatrician
- Viral hepatitis could be managed by GPs if they are confident to do so

To be undertaken by specialist practitioners (eg, Emergency Department / General Paediatric Team(s)):

- Treatment depends on the cause of hepatomegaly and needs input by the relevant specialist

When to Refer

See Suspected cancer: recognition and referral [NICE clinical guideline NG12]

URGENT referral to a paediatric specialist (irrespective of symptoms) is needed for:

- All infants with conjugated hyperbilirubinaemia
- All children with:
  - Significant cardiorespiratory symptoms
  - Haematemesis or melaena

  - Ascites (see Abdominal Distension)
  - Abnormal coagulation (eg, DIC)
  - Suspected liver mass
  - Blood film suggesting haematological malignancy
  - Altered consciousness

Non-urgent referral to a paediatric specialist:

- For all other cases (non-urgent, without 'red flag' symptoms or signs) of hepatomegaly, appropriate follow up should be arranged after discussing with the Paediatrician

'Safety Netting' Advice

- If condition worsens, inform medical practitioner

Resources

National Clinical Guidance

Suspected Cancer: recognition and referral (Web page), NICE clinical guideline NG12, National Institute of Clinical and Health Excellence.

Suggested Resources

Please note: these resources include links to external websites. These resources may not have national accreditation and therefore PCO UK cannot guarantee the accuracy of the content.

Guideline for the Investigation of Neonatal Conjugated Jaundice (PDF), British Society of Paediatric Gastroenterology, Hepatology and Nutrition.

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