

## Chapter 6: Gastroenterology and food allergy

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## **Chronic abdominal pain**

### **Definition**

It is intermittent or constant abdominal pain (either organic or functional) with induration of at least 2 months.

### **Risk factors/causes of functional abdominal pain**

- Genetic factors are thought to increase the likelihood that a child develops an FAPD
- Physical stressors in infancy and childhood predisposes the development of FAPDs later in childhood, adolescence and adulthood (e.g. history of cow's milk protein intolerance, allergic proctocolitis in infancy, gastrointestinal infection, Henoch-Schönlein purpura, abdominal surgery early in life, urinary tract infection in infancy)
- Psychosocial events and conditions throughout the patient's life (e.g. stressful life events, anxiety and depression)
- Psychological stressors early in life e.g. sexual, emotional, and physical abuse in childhood
- Parental deprivation such as unsatisfactory relationship between parents and losing a parent through death, divorce, or separation
- Consider gynaecological causes in girls who have achieved menarche

### **Promotion/prevention**

- Deworming
- Health Education

### **Signs and symptoms**

- Depends on the cause

### **Red flags in chronic abdominal pain**

#### Historical findings

- Involuntary weight loss (malabsorption)
- Unexplained fever
- Dysphagia, odynophagia
- Significant vomiting (protracted, bilious, projectile etc.)
- Chronic severe diarrhoea or nocturnal diarrhoea
- Urinary symptoms (change in bladder function, dysuria, haematuria, flank pain)
- Persistent right upper or right lower quadrant pain
- Back pain
- Family history of inflammatory bowel disease, coeliac disease, peptic ulcer disease
- Gastrointestinal blood loss (bloody diarrhoea/melena)
- Skin changes (rash, eczema, hives)
- Arthralgia

#### Examination findings

- Deceleration in linear growth
- Delayed puberty
- Oral aphthous ulcerations
- Abdominal tenderness
- Hepatomegaly and/or splenomegaly
- Costovertebral angle tenderness
- Perianal abnormalities (e.g. skin tags, fissures, fistulae)
- Abdominal mass
- Jaundice
- Pallor
- Psoriasis
- Signs of arthritis

### **Differential diagnosis**

#### Functional Abdominal Pain:

- Functional dyspepsia
- Functional abdominal pain not otherwise specified
- Irritable bowel syndrome
- Abdominal migraine

#### Organic causes:

- Peptic Ulcer disease

- Inflammatory bowel disease
- Coeliac disease
- Constipation
- Gastroesophageal reflux

### Rome IV diagnostic criteria for functional abdominal pain disorder

Disorder	Diagnostic criteria
<b>Functional dyspepsia (H2a)</b>	<p>Must include at least 1 of the following for at least 4 days per month:</p> <ul style="list-style-type: none"> <li>• Postprandial fullness</li> <li>• Early satiation</li> <li>• Epigastric pain or burning not associated with defecation</li> <li>• (Criteria must be fulfilled for at least 2 months.)</li> </ul> <p><b>H2a1. Postprandial Distress Syndrome</b> Includes bothersome postprandial fullness or early satiation which prevents finishing a regular meal. Supportive features include upper abdominal bloating, postprandial nausea, or excessive belching</p> <p><b>H2a2. Epigastric Pain Syndrome</b> Includes all of the following: bothersome (severe enough to interfere with normal activities) pain or burning localized to the epigastrium. The pain is not generalized or localized to other abdominal or chest regions and is not relieved by defecation or passage of flatus.</p> <p>Supportive criteria can include:</p> <p>(a) Burning quality of pain but without a retrosternal component</p> <p>(b) Commonly induced or relieved by ingestion of a meal but may occur while fasting</p>

<b>Irritable bowel syndrome - IBS (H2b)</b>	<p>Must include all of the following:</p> <ul style="list-style-type: none"> <li>• Abdominal pain at least 4 days per month associated with at least 1 of the following: related to defecation, a change in stool frequency, or a change in appearance of stool</li> <li>• In children with constipation, pain continues despite resolution of constipation (if pain resolves, the child has functional constipation)</li> </ul> <p>(Criteria must be fulfilled for at least 2 months)</p>
<b>Abdominal migraine (H2c)</b>	<p>Must include all of the following occurring at least twice:</p> <ul style="list-style-type: none"> <li>• Sudden episodes of intense, acute abdominal pain lasting at least 1 hour</li> <li>• Episodes are separated by weeks to months of mild or no abdominal pain</li> <li>• Typical pattern for each child</li> <li>• Pain is associated with at least 2 of the following: Anorexia, nausea, vomiting, headache, photophobia, or pallor</li> </ul> <p>(Criteria must be fulfilled for at least 6 months)</p>
<b>Functional abdominal pain— not otherwise specified - FAP-NOS (H2d)</b>	<p>Must include all of the following for at least 4 days per month:</p> <ul style="list-style-type: none"> <li>• Episodic or continuous abdominal pain not solely related to physiologic events (like eating or menses)</li> <li>• Does not meet criteria for other functional abdominal pain disorders (FAPD)</li> </ul> <p>(Criteria must be fulfilled for at least 2 months.)</p>

### Investigations

Investigate those with red flags based on underlying possible diagnosis.

- Full blood count
- Peripheral blood film
- ESR, CRP
- Liver Function Tests (LFT)

- Urea & electrolytes
- Urinalysis and culture
- Stool microscopy
- Stool occult blood testing
- Abdominal ultrasonography

### Management

- Treatment strategies need to be individualised based on the child's presentation

<b>Primary level</b> <ul style="list-style-type: none"><li>• Deworming therapy</li><li>• Refer all cases with red flags</li></ul>
<b>Secondary level</b> <ul style="list-style-type: none"><li>• Investigations as above</li><li>• Treat underlying causes</li><li>• Refer if cause not identified</li></ul>
<b>Tertiary level</b> <ul style="list-style-type: none"><li>• Investigate as above + other additional investigations as indicated</li><li>• Treat the underlying causes</li></ul>

### Follow up

- Nutrition and growth monitoring
- Follow up for underlying disease



## **Gastroesophageal Reflux Disease (GERD)**

### **Definition**

Gastroesophageal reflux (GER) refers to passage of gastric contents into the oesophagus.

GERD refers to reflux with pathological consequences such as oesophagitis, nutritional complications with weight loss or respiratory complications.

### **Causes/risk factors**

- Overweight, obesity
- Neurologic disorders (due to delayed gastric emptying)
- Cerebral palsy
- Hiatal hernia
- Asthma
- Stress
- Functional constipation
- Delayed gastric emptying
- Gastric outlet obstruction

### **Prevention/promotion**

- Caregivers and older children should be informed that excessive body weight is associated with an increased prevalence of GERD.
- Massage therapy, lifestyle interventions, or complementary treatments such as prebiotics, probiotics, or herbal medications should not be used to treat GERD.
- For patients on NGT feeding - education of parents on NGT care.
- For patients on gastrostomy feeding - education of parents on gastrostomy care.

### **Signs and symptoms General**

- Excessive irritability/pain
- Refusal of feeding
- Dental erosion
- Anaemia
- Weight loss or poor weight gain
- Dystonic neck posturing (Sandifer syndrome)

### **Gastrointestinal**

- Recurrent regurgitation with/without vomiting in the older child
- Heartburn/chest pain

- Epigastric pain
- Hematemesis
- Dysphagia/odynophagia
- Ruminant behaviour
- Esophagitis
- Oesophageal stricture
- Barrett oesophagus

### **Airway**

- Wheezing
- Stridor
- Cough
- Hoarseness
- Apnoea spells
- Apparent life-threatening events
- Asthma
- Recurrent pneumonia associated with aspiration
- Recurrent otitis media

### **Investigation**

- Full Blood count
- Urea and electrolytes
- Ultrasound
  - Look for signs of hiatal hernia, pyloric stenosis (first few months of age) and other disorders mimicking GERD
- Endoscopy
  - For direct visual examination and biopsy of the oesophageal mucosa to rule out macroscopic lesions such as esophagitis, erosions, exudate, ulcers, strictures, hiatal hernia, and others
- Barium studies
  - To diagnose anatomic abnormalities such as malrotation, duodenal web and stenosis; can also diagnose functional abnormalities such as achalasia

### **Differential diagnosis**

- Cow's milk protein allergy
- Eosinophilic esophagitis
- Foreign body ingestion
- Irritable bowel syndrome
- Oesophageal motility disorders
- Helicobacter pylori infection

- Hiatal hernia
- Intestinal motility disorders
- Other disorders associated with vomiting e.g. UTI, increased intracranial pressure, cyclic vomiting syndrome and metabolic disorders

## Management

### Primary level

- Physiologic GERD and regurgitation do not need medical treatment.
- Most will need parental reassurance, observation
- Lifestyle modifications
  - Weight loss
  - Avoid large meals (eat small frequent meals)
  - Wait 3 hours after a meal before lying down
  - Refrain from eating food (except liquids) within 3 hours of bedtime.
  - Elevate head of bed by 20°
- In infants, dietary treatment helps to decrease regurgitation. This includes:
  - Thickened formula and thickening agents
  - The addition of rice cereal (1 tablespoon) decreases the volume and frequency of regurgitation
  - Upright position after feeding
  - Advise against overfeeding babies
- If persists, refer to next level

### Secondary level

- As above
- Pharmacotherapy:
  - Antacids (not recommended for routine use in infantile GERD)
  - Proton pump inhibitor (omeprazole, esomeprazole): 1-2 mg/kg/day for 4 – 8 weeks as first line
  - If PPIs not available, use H<sub>2</sub> receptor antagonists (cimetidine)
- **Do not routinely use** prokinetic agents such as domperidone, metoclopramide, erythromycin, cisapride, and bethanechol as there is insufficient evidence to support their use
- The goal for medical therapy is to use the lowest doses for the shortest time possible

If it persists, refer to the next level

### **Tertiary level**

- As above
- Surgery indications include:
  - GERD with life-threatening complications such as apnoea or an apparent life-threatening event (ALTE) after failure of optimal medical treatment
  - Symptoms refractory to optimal therapy after appropriate evaluation
  - Chronic conditions (i.e. neurologic impairment, CP) with a significant risk of GERD- related complications
  - Need for chronic pharmacotherapy to control signs and/or symptoms of GERD beyond
  - the age of 2 to 3 years
  - Poor compliance to medication
  - Barrett's oesophagus

Refer to gastroenterologist

### **Follow up**

- Nutrition and growth
- Monitor complications of treatment

## **Peptic ulcer disease (PUD)**

### **Definition**

PUD is a spectrum of acid-related disorders that can affect the oesophagus, stomach and duodenum leading to mucosal barrier injury.

### **Risk factors/causes**

- Extreme physiologic stress, including trauma and sepsis
- Use of medications such as steroids, nonsteroidal anti-inflammatory drugs (NSAIDs)
- Hypersecretory state as in Zollinger-Ellison syndrome
- *H. pylori* infection
- Alcohol abuse (especially in the setting of psychiatric disorders)
- Smoking
- Psychological and work-related stress

### **Prevention/health promotion**

- Early diagnosis
- Avoid prolonged use of NSAIDs or steroids without health practitioner guidance
- Proton pump inhibitor use in cases of chronic use of NSAIDs or steroids
- Promote health literacy and key health messages

### **Signs and symptoms**

- Irritability
- Generalized abdominal pain
- Poorly localized abdominal pain (children will often have difficulty describing their symptoms)
- Epigastric abdominal pain
- Dyspepsia
- Vomiting
- Emesis
- Hematemesis
- Gastroesophageal reflux

**Red flags in PUD (should alert the physician that a child may truly have gastritis or PUD)**

- Hematemesis
- Involuntary weight loss
- Nocturnal awakening
- Poor appetite or early satiety
- Anaemia (iron deficient)
- Epigastric tenderness

**Investigations**

- FBC
- H. pylori test (stool antigen test or endoscopic biopsy with rapid urea testing)
- Endoscopy with biopsy
- Occult blood in stool examination

**Differential diagnosis**

- Oesophagitis
- Functional dyspepsia
- Gastritis
- Gastroenteritis
- Gastroesophageal disease

**Management**

**Primary level**

- Stabilize patients
- Discontinue aggravating medications e.g. NSAIDs, aspirin
- Administer antacids
- Refer all patients with suspected PUD to next level of care

**Secondary level**

- As above
- Treat underlying conditions
- H. Pylori eradication if confirmed
- Pharmacotherapy:
  - Antacids
  - Proton Pump Inhibitor (omeprazole, esomeprazole) in older children: 1-2 mg/ kg/day as first line
  - H2 receptor agonists (cimetidine)
- Refer all bleeding patients

**Tertiary level**

- Stabilize patient
- Investigations as above
- Pharmacotherapy as above
- Endoscopy in bleeding patients
- Gastroenterologist review

**Follow up**

- Follow up endoscopy if indicated.
- Confirmation of *Helicobacter pylori* eradication 4 weeks after eradication therapy.
- Medication and side effects.

## Chronic Diarrhoea

### Definition

Chronic diarrhoea is defined as three or more watery stools per day that persist for more than 2 weeks.

### Risk factors

- History of infectious diarrhoea
- Impaired immune function, primary or secondary immunodeficiency
- Underlying disease with faecal loading, such as Hirschsprung's disease
- Young age < 5 years
- Malnutrition
- Lack of breastfeeding
- Previous antibiotic use
- Food allergies

### Causes

#### Secretory diarrhoea

- Infections (rotavirus, adenovirus, cholera, entero-toxigenic *Escherichia coli* (ETEC), clostridium, giardia, cryptosporidium)
- Neuroendocrine disorders (Vasoactive intestinal peptide (VIP) secreting tumours e.g. neuroblastoma, pheochromocytoma)
- Endocrine disorders
- (hyperthyroidism, Addison's disease)
- Congenital diarrhoeas (congenital sodium diarrhoea, congenital chloride diarrhoea, Tufting enteropathy, Microvillus inclusion disease)

#### Malabsorption/maldigestion

- Infections (giardiasis, small intestinal bacterial overgrowth (SIBO), tropical sprue, Whipple disease)
- Galactosemia
- Ileal resection
- Exocrine pancreatic insufficiency e.g. cystic fibrosis
- Coeliac disease
- Causes of protein losing enteropathies (infections e.g. Tuberculosis (Tb), structural anomalies e.g. intestinal lymphangiectasia)

#### Inflammatory diarrhoea

- Infections (shigella, salmonella, E. coli, etc.)



- Inflammatory bowel disease (Ulcerative colitis, Crohn's disease)
- Food allergy

### Osmotic diarrhoea

- Lactose intolerance
- Osmotic laxatives
- Fructose intolerance

### Functional diarrhoeal disorders

- Irritable bowel syndrome
- Toddler diarrhoea

### **Prevention/promotion**

- Promote breastfeeding
- Water and sanitation hygiene (WASH)
- Vaccination
- Avoid unnecessary antibiotic use
- Restrict ingestion of large amounts of fruit juices

### **Signs and symptoms**

- Abdominal pain
- Weight loss/failure to thrive
- Fever
- Alternating diarrhoea and constipation
- Increased flatulence
- Abdominal bloating
- Other findings related to the specific aetiology

### **Investigations**

- Assess for malnutrition and other comorbid conditions e.g. Tb
- Stool studies
  - Stool microscopy (WBC, bacteria stain, parasite identification)
  - Stool culture
  - Perform faecal leukocyte, calprotectin, and lactoferrin studies and a Faecal Occult Blood Test
  - Consider further studies to help classify diarrhoea: e.g. faecal fat estimation, stool osmotic gap (differentiates osmotic from secretory diarrhoea. Stool osmolar gap <50 indicates secretory diarrhoea), stool pH (if < 6 suggests

carbohydrate malabsorption)

- Stool alpha 1 antitrypsin >0.5mg/g suggests a protein losing enteropathy
- Stool elastase level <200mcg/g suggest pancreatic insufficiency (note loose stool may have a dilution effect and give a false low elastase level)
- Blood tests:
  - HIV test
  - FBC & differential count & diagnostic studies for anaemia, acanthocytes on peripheral smear may suggest fat malabsorption
  - Serum electrolytes including calcium, magnesium, phosphates, liver blood profile
  - ESR/CRP
  - Blood gas, random blood sugar
- Imaging studies - Use imaging studies to evaluate red flags in diarrhoea and consider initially to rule out structural disease.
  - CT abdomen or MRI abdomen with enterography
- Endoscopy/colonoscopy with biopsy for suspect inflammatory bowel disease, coeliac disease and congenital diarrhoeas
- Other investigations specific to the suspected aetiology, including immunoglobulin levels (IgA, IgG, IgM as basic screen for primary immune deficiencies)

## Management

<b>Primary level</b> <ul style="list-style-type: none"> <li>Correct dehydration and refer</li> </ul>
<b>Secondary</b> <ul style="list-style-type: none"> <li>Investigations as above</li> <li>Correct dehydration</li> <li>Treat infectious causes/malnutrition</li> </ul> <p>Refer if diarrhoea persists or non-infectious aetiology identified</p>
<b>Tertiary</b> <ul style="list-style-type: none"> <li>Investigate as above</li> <li>Correct dehydration and electrolyte imbalances</li> <li>Treat the underlying causes</li> </ul> <p>Consult gastroenterologist for complex conditions</p>

**Follow up**

- Nutrition and growth
- Refer to relevant clinics

## Constipation

### Definition

Constipation is a delay or difficulty in defecation, for 2 or more weeks, which is sufficient to cause significant distress to the patient.

### Risk Factors/causes

Usually multifactorial:

- Includes environmental conditions e.g. diet
- Physical disabilities, e.g. cerebral palsy
- Painful or frightening defecation
- Age difference risk factors are as below:

#### Toddlers

- Dietary changes (breastmilk to formula or cow's milk) lead to dry hard stools with fissures and pain.
- Toilet training: Excessive parental pressure, anxiety

#### Older children

- Unpleasant toilet facilities away from home
- Sexual abuse
- Trauma to the perianal area
- Voluntary withholding while playing

### Prevention and Promotion

- Encourage a high fibre diet, including beans, vegetables, fruits, whole grain cereals.
- Encourage the child to eat fewer foods with low fibre such as processed food.
- Encourage the child to drink plenty of fluids.
- Encourage the child to stay active and get regular exercise.
- Schedule time for toilet visits.

### Signs and symptoms

- A history and physical examination are usually sufficient to distinguish functional constipation from constipation caused by an organic condition.
- The Rome IV criteria are the most accepted criteria for diagnosing childhood constipation.

**Rome IV diagnostic criteria for diagnosing functional constipation in children****At least two of the following in a child with a developmental age younger than four years\***

- Two or fewer bowel movements per week
- At least one episode of incontinence per week
- At least one episode of incontinence per week after the acquisition of toileting skills
- History of excessive stool retention
- History of painful or hard bowel movements
- Presence of a large faecal mass in the rectum
- History of large diameter of stools that may obstruct the toilet

**At least two of the following in a child with a developmental age of 4 years or older with insufficient criteria for irritable bowel syndrome<sup>#</sup>**

- Two or fewer bowel movements in the toilet per week
- At least one episode of faecal incontinence per week
- History of retentive posturing or excessive voluntary stool retention
- History of painful bowel movement
- Presence of a large stool mass in the rectum
- History of large diameter of stool that may obstruct the toilet

\*Criteria must be fulfilled for at least 1 month.

<sup>#</sup> Criteria must be fulfilled at least once a week for at least 1 month

Further evaluation may be warranted in children with red flags that might suggest an organic aetiology as below:

Red Flags	Suggested diagnoses
Age of onset $\leq$ 1 month	Anorectal malformation or spine malformation, Hirschsprung disease (HD), allergy, metabolic/ endocrine condition
Delayed passage of meconium more than 48hrs after birth	HD, cystic fibrosis, anorectal malformation, spine malformation, congenital hypothyroidism
Failure to Thrive	HD, malabsorption, cystic fibrosis, metabolic condition

Abdominal distention	HD, impaction
Sacral dimple covered by a tuft of hair / abnormal neurological exam	Spinal cord abnormality
Extra-intestinal symptoms like vomiting, and ill appearance, anaemia, jaundice, clubbing, respiratory problems, joint disease	May indicate an organic cause, e.g. HD, Coeliac ds
Gushing of stool with rectal examination	HD
No response to conventional treatment	HD, spinal cord problem

### Investigations

- Review growth chart to assess growth
- FBC
- TSH and T4 can screen for hypothyroidism
- Contrast enema to evaluate for Hirschsprung disease (HD)
- Abdominal ultrasonography (if not available then X-ray abdomen) to show faecal impaction (in a child whose abdominal examination is difficult but is not done routinely)
- Magnetic resonance imaging of the spine may be necessary to evaluate for a tethered cord, spinal cord tumour, or sacral agenesis.
- Rectal biopsy

### Differential Diagnosis

- Functional constipation
- HD
- Anatomical malformation (colon/rectal stenosis/ imperforate anus)
- Spinal cord abnormalities (MMC)
- Cerebral palsy
- Hypothyroidism
- Hypercalcemia, hypokalaemia, other electrolyte abnormalities
- Breast fed children or formula fed children
- Drugs e.g. opioids, phenobarbital
- Cow's milk protein intolerance
- Coeliac disease
- Inflammatory bowel disease

**Management****Primary level**

- Health promotion steps
- Patients with red flags or suspected organic causes should be referred to the secondary and tertiary level of care

**Secondary level**

See the tertiary-level guidance below

**Tertiary level**

- Algorithms for the evaluation and management of constipation in infants and older children are presented in Figures 1 and 2.

**Disimpaction (enema)**

- The first phase of treatment is to empty the hard stool from the colon, also known as disimpaction.
- In the past, manual removal, suppositories, and enemas were common methods during this phase of treatment. Polyethylene glycol (PEG 3350) has become the first treatment of functional constipation due to its efficacy, safety profile, and because it is well tolerated.
- A reasonable dose is 1 to 1.5 grams per kilogram PEG 3350 mixed with 200 mL water or juice.
- Patients should be encouraged to drink this over 3 hours, if possible.
- If there has not been a significant response to this treatment, the patient can repeat the dose the next day.
- If there is no response after two days of treatment or significant abdominal discomfort, persistent vomiting, or any other concerns, the family should present for follow-up and re-evaluation.
- Below is a summary of therapies for dis-impaction in children:

**Therapies for dis-impaction in children**

THERAPY	DOSE
<b>Osmotics</b>	
Polyethylene glycol 3350	1.5g/kg/day
Lactulose	1-3 ml/kg/day of 3.3g/5ml solution
<b>Stimulants</b>	
Bisacodyl	Over 2 years: 5 – 15 mg (1 – 3 tablets) per day in single dose
<b>Enema (one per day)</b>	
Saline	5 – 10 mL per kg
Mineral oil	15 – 30 mL per year of age up to 240 mL
<b>Suppository (one per day)</b>	
Bisacodyl	≥ 2 years: 5 to 10 mg (½ to 1 suppository)
Glycerine	½ to 1 infant suppository Adult suppository for those older than 6 years

**Maintenance Therapy**

- In the second phase of treatment, the goal is to keep the stool very soft,
  - preventing re- accumulation of hard stool while the colon returns to normal
  - size and function. Drugs in this phase are oral medications.
  - Osmotic laxatives
    - Polyethylene glycol (PEG) 3350 at 0.2-0.8 g/Kg/day
    - Lactulose at 1- 3 mL /kg/day
    - Magnesium hydroxide at 0.5-3 mL/kg/day
  - Stool Softeners
    - Docusate sodium at 5 mg/kg/day
    - Mineral oil (lubricant) at 1-3 mL/Kg/day
  - Stimulant laxative for rescue therapy in addition or alone (duration less than 30 days)
    - Senna at 2.5-7.5 mL/day
    - Bisacodyl at 5-10 mg/day
- Non-Pharmacological - Same as preventative measures outlined above.

**Follow up**

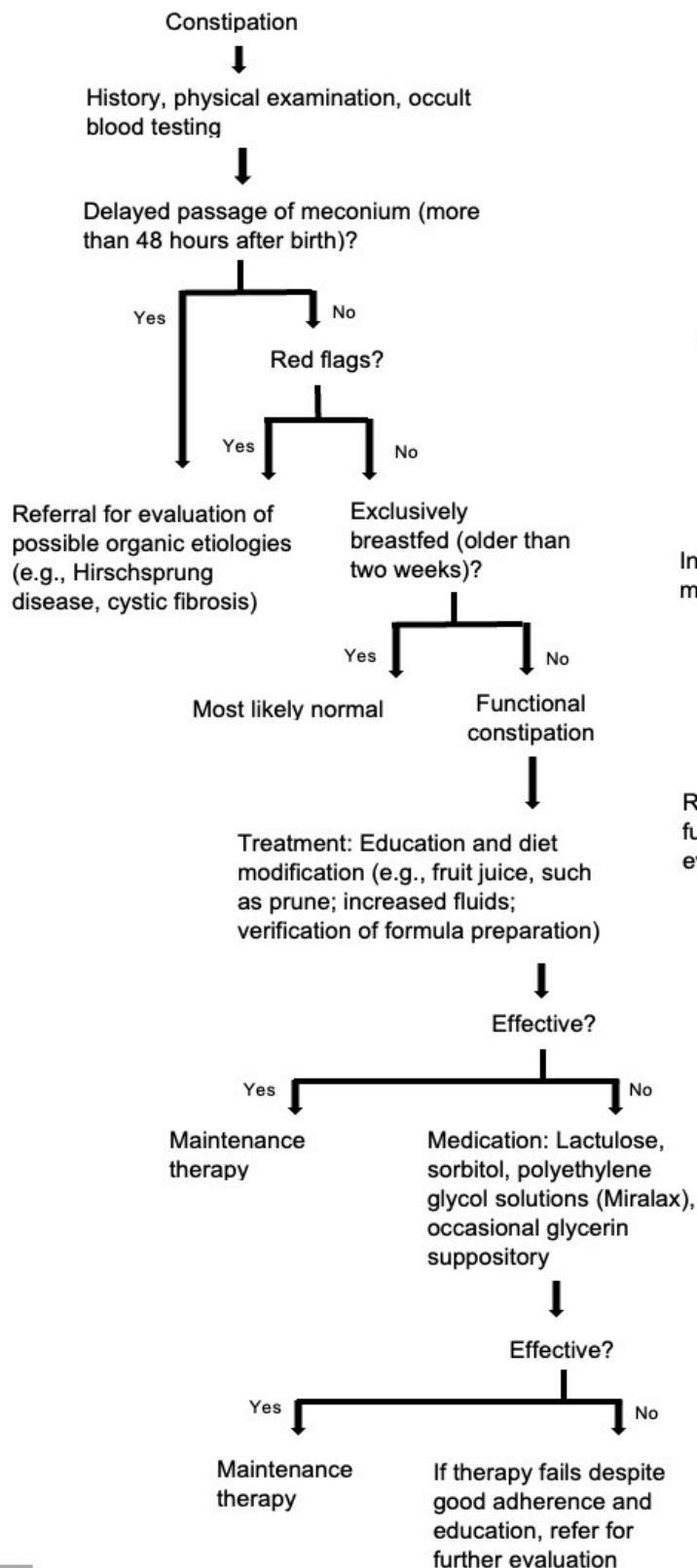
- Assessing for complications



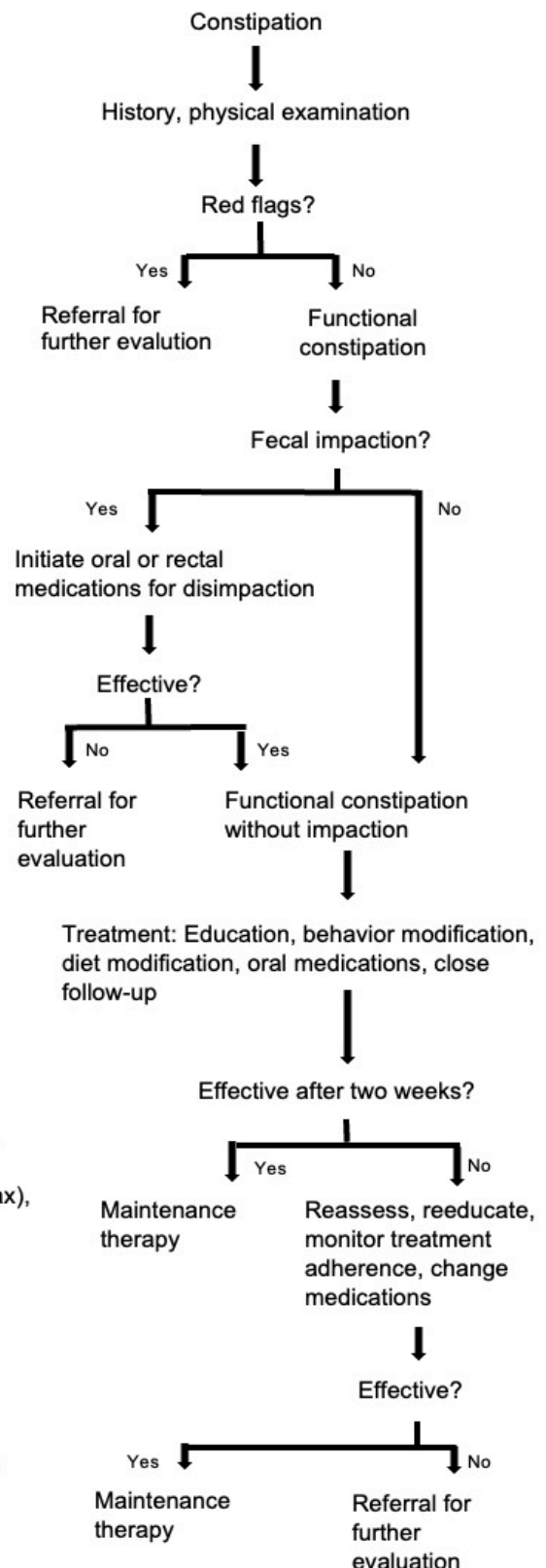
## Chapter 6: Gastroenterology and food allergy

- Medication side effects
- Growth and development

**Constipation in infants younger than six months**



**Constipation in children six months and older**



## Food Allergy

### Definition

Food allergy is an adverse health effect arising from a specific immune response that occurs reproducibly on exposure to a given food. Can be:

- IgE mediated Type 1 hypersensitivity reaction (immediate onset within minutes - 2hrs of ingestion)
- or
- Mixed IgE/non-IgE mediated reactions (delayed onset hours to days after ingestion)

Food intolerance is an adverse reaction to food or a food component that lacks an identified immunologic pathophysiology.

### Risk factors/causes

- Peanuts / tree nuts
- Fish / Seafood
- Milk (bovine / soy)
- Eggs
- Seeds (maize) and other foods
- Fruits
- Positive Family history

### Prevention/promotion

- Education of children, parents and carers is the mainstay of dietary avoidance advice. Educate on:
  - Avoidance of known food allergens by checking common ingredients and reading food labels
  - Appropriate safe, cost-effective, freely available and nutritionally adequate substitutes for avoided foods
- Encourage exclusive breastfeeding for the first 4-6 months
- Early introduction of complementary foods (including potential allergens) by 4-6 months of age (not applicable to infants experiencing allergic reactions)
- Health talks and increased awareness
- Encourage susceptible children not to eat food which is not prepared by a person who is aware of their conditions
- Advocate for availability of epi-pens and teach the parents/child/caregiver how to administer

## Signs and symptoms

### IgE mediated food allergy

- Symptoms usually recur on exposure to the food on every occasion. They may be mild or severe, associated with anaphylaxis.
  - Skin (most common): pruritus, urticaria, atopic dermatitis, exanthem, angioedema
  - Respiratory: rhinitis, sneezing, nasal congestion, dyspnoea, wheezing, laryngeal oedema (can be fatal)
  - Gastrointestinal: nausea, vomiting, abdominal pain, diarrhoea, oral itching/swelling
  - Cardiovascular: hypotension, tachycardia, dysrhythmias
  - CNS: Headache

### Non-IgE/mixed food allergy

- Symptoms typically limited to skin and gastrointestinal tract.
  - Skin: atopic dermatitis
  - Gastrointestinal: abdominal pain, vomiting, diarrhoea, bloody stools, failure to thrive, eosinophilic oesophagitis, food protein-induced enterocolitis syndrome, allergic proctocolitis and enteropathy syndromes
- A clear cause-effect relationship between exposure to the suspected food and symptoms is not always possible, as symptoms develop over time and are more chronic in nature

## Investigations

- Patient History **vital** in establishing diagnosis
- FBC
- Suspected IgE mediated food allergies
  - Skin prick testing
  - Serum specific IgE antibodies to suspected foods are used to prove sensitization. Diagnosis requires a clear correlation between the test result and clinical reaction (by positive history or food challenge)
  - Total IgE antibody serum levels
  - If above tests are inconclusive try elimination diet: the suspected allergens are eliminated from the patient diet while being observed for an improvement in symptoms without the need for medication

### Non IgE mediated food allergies

- No validated tests exist to confirm non-IgE- or mixed IgE/non-IgE-mediated food allergies.
- In certain cases, endoscopy with biopsy is indicated to evaluate the response to

dietary changes.

### Differential diagnosis

- Psychological reactions (food aversion)
- Organic reactions (e.g. peptic ulcer disease)
- Anatomical reactions (e.g. strictures)
- Toxic reactions (e.g. food poisoning)
- Non-toxic reactions

### Management

<b>Primary level</b> See the secondary-level guidance below
<b>Secondary level</b> <ul style="list-style-type: none"> <li>• ABCDE approach if acutely unstable</li> <li>• Manage anaphylaxis (refer to section in emergency chapter)</li> <li>• Perform thorough history</li> </ul> Refer all patients with suspected food allergy
<b>Tertiary level</b> <ul style="list-style-type: none"> <li>• ABCDE approach if acutely unstable</li> <li>• Manage anaphylaxis (refer to section in emergency chapter)</li> <li>• Take thorough history</li> <li>• Eliminate offending food from diet</li> <li>• Provide with auto injectable epinephrine (EPI-PEN) for emergency home treatment                         <ul style="list-style-type: none"> <li>• 8-25 kg 0.15mg/dose</li> <li>• &gt;25 kg 0.3mg/dose</li> </ul> </li> <li>• Refer to dietician/allergologist/gastroenterologist for further management tailored to each child</li> </ul>

### Follow up

- Continued counselling and education
  - Severity of future allergic reactions is not accurately predicted by past history or allergy test results
- Refill of EpiPen and continued family training
- Provision of individualised written instructions/care plan on the indications for and method of administration of emergency medication
- The diagnosis of food allergy should, with permission, be communicated to all

relevant caregivers, including school teachers

- Patients should be encouraged to join an appropriate patient support organisation

## Inflammatory bowel disease

### Definition

- Inflammatory bowel diseases (IBD) are a spectrum of diseases characterised by recurrent inflammation of the intestine
- Can be categorised into three main subtypes:
  - Crohn's Disease is characterised by transmural, granulomatous inflammation affecting any part of the gastrointestinal tract from the mouth to the anus, often discontinuously.
  - Ulcerative Colitis (UC) is limited to the colon and consists of superficial ulceration of the bowel mucosa.
  - Inflammatory bowel disease unclassified (IBDU) describes patients with chronic colitis within the spectrum of IBD but in the absence of distinguishing features of either CD or UC.
  - <10 years: early onset IBD
  - <6 years: very early onset IBD (VEO-IBD) – higher likelihood of underlying monogenic aetiology or primary immune deficiency

### Risk factors/causes

- Low-fibre, high-sugar diets
- Sedentary lifestyle
- Diets poor in fruits and vegetables and high in animal fats and sugar
- Emulsifiers commonly found in processed food
- Antibiotic use in early childhood and dysbiosis of the gut microbiota
- Vitamin D deficiency
- Intestinal infections
- Stress
- Sleep deprivation
- Other autoimmune diseases e.g. enthesitis arthritis
- Genetics - having a first- or second-degree relative with IBD
  - Onset below 6 years of age

### Prevention/promotion

- Health Education
- Over-the-counter and prescription medicines should be avoided
- Avoid taking ibuprofen or other NSAIDs
- Avoid unnecessary antibiotic use

**Signs and symptoms Intestinal Manifestations:**

- Abdominal pain
- Abdominal distension (UC > CD)
- Right iliac fossa mass (CD)
- Diarrhoea  $\pm$  blood/mucus; urgency and tenesmus (proctitis)
- Rectal bleeding
- Perirectal disease, fistula
- Nausea/vomiting
- Anorexia; weight loss; lethargy (CD > UC)
- Aphthous oral ulcers (CD > UC)
- Fever
- Growth retardation (CD > > > UC)
  - May begin before the development of specific gastrointestinal symptoms
  - May be the only presenting clinical symptom

**Extraintestinal manifestations:**

- Arthralgia, arthritis, ankylosing spondylitis
- Enthesitis, myositis, erythema nodosum, pyoderma gangrenosum
- Uveitis, episcleritis, iritis, conjunctivitis
- Finger clubbing
- Anaemia
- Thromboembolism, vasculitis
- Urinary tract obstruction, renal stones
- Delayed puberty, hepatic disease
- Nutritional deficiencies: iron, vitamin D, vitamin B<sub>12</sub> and folic acid deficiency
- Depression and anxiety
- Hepatobiliary manifestations: primary sclerosing cholangitis

**Differential diagnosis of presenting symptoms of inflammatory bowel disease**

Primary presenting symptom	Diagnostic consideration
Right lower quadrant abdominal pain, with or without mass	Appendicitis, abscess, infection (e.g., <i>Campylobacter</i> , <i>Yersinia</i> ), lymphoma, intussusception, mesenteric adenitis, Meckel diverticulum, ovarian cyst
Chronic periumbilical or epigastric abdominal pain	Irritable bowel syndrome, constipation, lactose intolerance, peptic ulcer disease, coeliac disease
Rectal bleeding, no diarrhoea	Fissure, polyp, Meckel diverticulum, rectal ulcer syndrome, vascular abnormalities



Bloody diarrhoea	Infection (intestinal TB, amoeba, CMV), haemolytic-uremic syndrome, Henoch-Schönlein purpura, cow milk protein allergy
Watery diarrhoea	Irritable bowel, lactose intolerance, giardiasis, cryptosporidium, sorbitol, laxatives
Perirectal disease	Fissure, haemorrhoid (rare), streptococcal infection, condyloma (rare)
Growth delay	Coeliac disease, endocrinopathy
Anorexia, weight loss	Anorexia nervosa
Arthritis	Collagen vascular disease, infection
Liver abnormalities	Chronic hepatitis (Infection, Autoimmune hepatitis, Coeliac disease, Wilsons Disease)

### Investigations/evaluation

#### Urinalysis

#### Blood tests

- Full blood count
- ESR/CRP
- ALT, AST, GGT
- Total protein/Albumin

#### Stool tests

- Microscopy, culture and sensitivity
- Stool for occult blood
- Stool calprotectin or lactoferrin

#### TB screening tests

#### Imaging (for localization of small bowel disease)

- Upper GI series/small bowel follow through

- Abdominal CT with oral contrast
- Magnetic resonance enterography (MRE)

#### Endoscopy

- Ileo-colonoscopy with biopsies
- Upper endoscopy with biopsies

#### Specialised tests depending on clinical findings:

- Perinuclear antineutrophil cytoplasmic antibody (P-ANCA) - UC
- anti-saccharomyces cerevisiae (ASCA) - CD
- Lactose/glucose hydrogen breath test
- 72hours faecal fat quantification
- Stool alpha-1 antitrypsin
- Cholinesterase
- Serum iron, calcium, magnesium, folate, zinc, vitamins A/E/B12

### Management

<b>Primary level</b> Refer suspected cases
<b>Secondary level</b> Refer suspected cases
<b>Tertiary level</b> <ul style="list-style-type: none"> <li>• Supportive treatment</li> <li>• If severely ill, keep nil by mouth with IV hydration +/- parenteral nutrition</li> <li>• Provide adequate pain relief</li> <li>• Pharmacologic treatment <ul style="list-style-type: none"> <li>• Oral 3-aminosalicylic acid (ASA) dimers (mesalazine and sulfasalazine) <ul style="list-style-type: none"> <li>◦ Can be used to induce and maintain mild-moderate CD colonic inflammation.</li> </ul> </li> <li>• Corticosteroids (induction therapy)</li> <li>• Oral prednisone 1 mg/kg daily (maximum of 40 mg but up to 60 mg/day), intravenous therapy for severe disease: methylprednisolone at 1 to 1.5 mg/kg (maximum daily dose of 60 mg)</li> </ul> </li> </ul> <p><b>OR</b></p> <ul style="list-style-type: none"> <li>• Budesonide 9 mg/day (Compared with prednisone, it has fewer corticosteroid side effects so can be used for a longer period)</li> <li>• For 6 weeks tapered over 2 weeks for the treatment of ileocecal disease.</li> <li>• Corticosteroid enemas can be used to treat rectal disease.</li> <li>• Glucocorticoids are <b>not</b> effective as maintenance therapy and are</li> </ul>

associated with significant side effects.

- Biological Therapy
- Anti-TNF $\alpha$  antibodies (Infliximab, Adalimumab, Certolizumab). Indications are:
  - Moderate-to-severe luminal CD
  - Corticosteroid-dependent or corticosteroid-refractory disease
  - Failure of response to immunomodulators
  - Fistulizing disease, especially perianal fistulas
- Superior outcomes may be achieved if used within 3 months of diagnosis
- Antibiotics (Ciprofloxacin or metronidazole may be helpful, especially in fistula disease)
- Dietary treatment
- Modular/elemental diets to induce remission (CD > UC)
- Dietary supplementation to minimise poor growth and correct specific nutritional deficiencies: vitamin D, vitamin B12, folic acid, mineral supplements like iron

Consult gastroenterologist/dietician/surgeon

### Follow up

- Adherence to the prescribed medications for IBD is critical for disease control
- Advise on appropriate diet
- Patients should drink enough fluids to be well hydrated
- If patients are on steroids, they should get plenty of calcium in the diet to maintain healthy bones. Consider taking a calcium supplement with vitamin D
- Patients should keep a food diary to identify foods that make symptoms better or worse, and avoid foods that cause symptoms

## **Coeliac disease**

### **Definition**

Coeliac disease (CD) is an immune-mediated enteropathy due to intolerance to gluten protein (present in wheat, barley and rye).

### **Causes/risk factors**

- Consumption of wheat, rye and barley-containing grains by genetically susceptible individuals
- Genetic factors: increased incidence in girls, higher prevalence of CD among first-degree relatives (occurrence of multiple cases in families)
- Associated conditions: Down syndrome, insulin-dependent type 1 diabetes mellitus, Hashimoto thyroiditis, Addison disease, selective IgA deficiency

### **Prevention and promotion**

- Avoid gluten exposure
- Family members should get screened for early signs of CD
- Educate on appropriate diet

### **Clinical features**

- Failure to thrive
- Chronic diarrhoea
- Pale, bulky, floating stools
- Constipation
- Abdominal distension
- Vomiting
- Anorexia
- Irritability
- Later manifestations
  - Anaemia (iron deficiency)
  - Apathy
  - Ascites
  - Peripheral oedema
  - Short stature
  - Delayed puberty
  - Arthralgia
  - Hypotonia, muscle wasting
  - Specific nutritional disorders (vitamin D, iron)
  - Coeliac crisis: Life threatening dehydration due to diarrhoea accompanying

malabsorption

## Investigations

Diagnosis is confirmed by positive tissue transglutaminase IgA antibodies + positive mucosal histology and full recovery on gluten-free diet

### Serum

- Antiendomysial antibodies e.g. tissue transglutaminase and antireticulin IgA antibodies (beware of false negatives in IgA deficiency)
- Total IgA
- Antigliadin antibody

### Endoscopy with biopsy of duodenum

- Shows lymphocyte infiltration, diffuse subtotal villous atrophy, crypt hyperplasia

## Differential diagnosis

- Transient gluten intolerance post gastroenteritis
- Cow's milk protein intolerance
- Giardiasis
- Crohn's disease

## Management

<b>Primary level</b> Manage dehydration and refer
<b>Secondary level</b> See the tertiary-level guidance below
<b>Tertiary level</b> <ul style="list-style-type: none"> <li>• Supportive management <ul style="list-style-type: none"> <li>• Treat life threatening dehydration or anaemia (refer to emergency section)</li> </ul> </li> <li>• Gluten-free diet under supervision of a dietician <ul style="list-style-type: none"> <li>• Rice, maize, and buckwheat are nontoxic and are usually used as wheat substitutes. Chestnut, cassava, sorghum, millet, teff, quinoa, and amaranth are other safe starchy foods.</li> <li>• Gluten avoidance should be lifelong.</li> </ul> </li> <li>• Nutritional supplements (vitamin D, iron) may be required</li> <li>• Consult Gastroenterologist</li> </ul>

## Acute liver failure

### Definition

Coagulopathy with INR  $\geq 1.5$  with encephalopathy or INR  $\geq 2$  without encephalopathy due to a liver cause, not correctable by intravenous vitamin K, with biochemical evidence of acute liver injury and no evidence of chronic liver disease

### Subtypes:

- Hyperacute liver failure (0 - 1 week)
- Acute (fulminant) liver failure (8 - 28 days)
- Subacute liver failure (4 - 12 weeks)

### Risk factors/causes

- Idiopathic (20-45% of cases)
- Viral Infections (Hepatitis A/B/B+D/E, CMV, EBV, HSV, Adenovirus, varicella, measles)
- Non-viral infections (Salmonella, TB, Malaria, gram negative sepsis, Toxoplasmosis)
- Hepatotoxic medications
  - Acetaminophen
  - Antimicrobials (amoxicillin, ciprofloxacin, cephalosporins, TB treatment, Ketoconazole, antiretrovirals)
  - Anticonvulsants (phenytoin, carbamazepine, valproic acid)
  - Chemotherapy
- Other toxins (herbal supplements, alcohol, aflatoxins, cocaine, mushroom poisoning)
- Malignant infiltration (HLH, Leukaemia, lymphoma)
- Ischaemia
  - Acute circulatory failure (shock, cardiac failure, myocarditis)
  - Tissue hypoxia due to respiratory failure
  - Budd-Chiari syndrome
  - Ischemic hepatitis
- Genetic/metabolic (galactosaemia, Wilson disease, inborn urea cycle defects)
- Autoimmune hepatitis
- Gestational Alloimmune liver disease (neonatal acute liver failure, with haemochromatosis)

### Signs and symptoms

Presentation mostly non-specific and clinicians should maintain a high level of suspicion. Signs/ symptoms include:

- Fatigue, lethargy, malaise
- Jaundice
- Anorexia
- Nausea, vomiting
- Abdominal pain (RUQ pain or generalised pain)
- Signs of hepatic encephalopathy
  - Altered level of consciousness
  - Asterixis
- Symptoms of cerebral oedema
- Pruritus
- Features of underlying aetiology

### **Health promotion/prevention**

- Vaccination
- Educate on danger of herbal remedies
- Drug and toxin storage in appropriate places away from children
- Careful medication use in communities and hospitals
- Educate on danger of alcohol abuse
- Education on appropriate storage of groundnuts and maize for consumption to reduce aflatoxin exposure

### **Investigations**

#### Blood tests

- Random blood sugar
- HIV test
- Malaria screen
- Full blood count
- CRP
- Liver chemistries - ALT, AST, GGT, ALP, Bilirubin, Albumin, Total protein
- Coagulation Tests - PT/INR
- Serum Electrolytes, Urea, Creatinine
- Blood gas with lactate
- Serum ammonia
- Hepatitis A, B, C, E, HSV, Syphilis
- Toxicology screen (e.g. acetaminophen levels)
- Other tests if indicated:
  - Older children: autoimmune screen and Wilson Diseases screen (ceruloplasmin)
  - Ferritin level and transferrin saturations, triglycerides and metabolic screen-urine reducing substances, urine organic acids, serum amino acids (In

babies <6 months with liver failure)

Imaging (findings vary as liver failure progresses)

- Abdominal USS
  - Heterogeneously decreased liver echogenicity (liver necrosis), ascites
  - In patients who have been sick for at least 7 days, Ultrasound may show a nodular surface which can be mistaken for cirrhosis
- Doppler ultrasound - to identify underlying cause e.g. portal vein thrombosis/hepatic ischemia

Liver biopsy indications

- To distinguish between acute liver failure and chronic liver disease if the diagnosis is uncertain
- If the suspected underlying aetiology requires specific management e.g. autoimmune hepatitis
- Other tests for specific aetiologies can be done based on clinical suspicion

### Differential diagnosis

- Other encephalitis or encephalopathy e.g. drugs and toxins, uremic encephalopathy
- Sepsis
- Other coagulation disorders

### Management

#### Primary level

- Emergency Stabilisation ABCDE approach
  - Volume repletion (Normal Saline 20ml/kg bolus)
  - Treat hypoglycaemia
- Refer

#### Secondary level

- ABCDE approach
- Aggressive supportive care
  - Volume repletion (normal saline 20ml/kg bolus - repeat if necessary)
  - Treat hypoglycaemia
  - Treat electrolyte disturbances
  - Optimise Nutrition (maintain 1-2 mg/kg/day protein intake)
  - Stress ulcer prophylaxis to prevent GI bleeding
- Control Hyperammonaemia
  - Lactulose 0.5ml/kg/dose - titrate to produce 2-4 loose stools daily



- Neuroprotective measures
  - Elevate head of bed to 30°
  - Maintenance of normoxia, normocapnia, normotension, normothermia, euglycemia
- Correct coagulopathy
  - IV Vitamin K (300 mcg/kg/day, [max 10mg] for at least 3 days)
  - FFP transfusion (15ml/kg) recommended if active bleeding
- Infection control - antibiotic prophylaxis (ceftriaxone 50 mg/kg/day)

Refer to tertiary facility

### **Tertiary level**

- ABCDE approach
  - Early intubation in patients with rapidly progressing encephalopathy
- Aggressive supportive care
  - Volume repletion (Normal Saline 20mls/kg bolus - repeat if necessary)
    - Start vasopressors if not responsive to fluids
  - Treat hypoglycaemia
  - Treat electrolyte disturbances
  - Optimise Nutrition (maintain 1-2 mg/kg/day protein intake)
  - Stress ulcer prophylaxis to prevent GI bleeding
- Neuroprotective measures
  - Maintenance of normoxia, normocapnia, normotension, normothermia, euglycemia
  - Elevate head of bed to 30°
- Manage high ICP
  - Mannitol (0.5 - 1g /kg) IV over 30 minutes. Repeat 8hrly.
  - 3% Hypertonic saline (0.5-3ml/kg/hr)
- Control Hyperammonaemia
  - Lactulose 0.5ml/kg/dose - titrate to produce 2-4 loose stools daily
- Correct coagulopathy
  - IV Vitamin K (300 mcg/kg/day, [maximum 10mg] for at least 3 days)
  - FFP transfusion (15ml/kg) if bleeding actively
- Infection control - antibiotic prophylaxis (ceftriaxone 50 mg/kg/day)
- Manage kidney injury
  - Optimise hemodynamic support
  - Renal replacement therapy if necessary
- N-acetylcysteine in acetaminophen toxicity and when cause unknown
- Treat underlying cause

**Follow up**

- Check for progress of disease/complications

## **Liver Cirrhosis**

### **Definition**

Liver cirrhosis is the advanced stage of liver fibrosis and is the common endpoint of many different liver diseases

### **Risk factors/causes**

- Cholestatic liver disease e.g. biliary atresia, choledochal cyst
- Metabolic liver disease e.g. Wilsons' disease, glycogen storage disease
- Vascular disease e.g. heart failure
- Drugs
- Herbal medication
- Toxins
- Infections
- Autoimmune hepatitis

### **Promotion/prevention**

- Avoid exposure to risk factors that are associated with acute liver injury/disease
- Encourage vaccination according to Extended Programme on Immunization (EPI)
- Routine screening of pregnant women for Hepatitis B and appropriate treatment of exposed babies
- Alcohol education for adolescents
- Early referral of infants with congenital liver diseases

### **Signs and symptoms**

- Patients initially asymptomatic
- Fatigue, malaise, anorexia
- Weight loss/malnutrition/signs of micronutrient deficiency (fat soluble vitamins A, D, E, K)
- Peripheral oedema/ anasarca
- Feter hepaticus
- Abdominal features (Jaundice, nausea, vomiting, abdominal distension, hepatomegaly, splenomegaly, ascites, pale stools, dark urine) - liver can be shrunken, small and impalpable in advanced cirrhosis
- Skin features (spider naevi, palmar erythema and prominent periumbilical veins, caput medusae, petechiae, purpura, dry and atrophic skin, scratch marks)
- Finger clubbing, leukonychia, asterixis
- Variceal bleed with hematemesis, coffee ground vomitus, and/or melena
- Recurrent epistaxis and spontaneous bruising/bleeding

- Signs of hyperoestrogenism
  - Males - gynaecomastia, hypogonadism, feminisation
  - Females – amenorrhoea

## Investigations

### Blood tests

- Random blood sugar
- HIV test
- Full blood count and grouping and cross matching if indicated
- Liver chemistries - ALT, AST, GGT, ALP, Bilirubin, Albumin, Total protein
- Coagulation Tests - PT / INR
- Serum Electrolytes including calcium, magnesium, phosphate
- Urea, Creatinine
- Hepatitis B, C, Tb, Syphilis, Schistosomiasis
- Toxicology screen (e.g. acetaminophen levels)
- Alpha-fetoprotein (AFP), LDH

### Imaging (findings vary as liver failure progresses)

- Abdominal USS with doppler
  - Heterogeneous nodular liver with fibrous septa. May be enlarged and shrunken, portal hypertension, ascites, splenomegaly, increased portosystemic collateral flow.
- Cardiac echo to exclude cirrhotic cardiomyopathy
- Plain radiographs of the wrist and knee to demonstrate bone age and/or the development of osteopenia or osteomalacia (rickets/hepatic osteodystrophy)

### Endoscopy

- Upper GI Endoscopy to identify oesophageal varices and peptic ulcer
- Lower GI Endoscopy to identify source of bleeding

### Liver biopsy (gold standard). Indications are:

- In cases of uncertain diagnosis
- Grading and staging of inflammation and fibrosis
- Monitoring treatment response e.g. in autoimmune hepatitis
- Evaluation of focal lesions

Other tests for specific aetiologies can be done based on clinical suspicion

**Differential diagnosis**

See section on risk factors and causes

**Management****Primary level**

See secondary-level guidance. below

**Secondary level**

- Stabilise the patient
- Check RBS and correct any hypoglycaemia
- Manage bleeding with Vitamin K (2.5–10 mg/day)
- Nutritional assessment (mid-arm circumference) and provide nutritional support
- Involve palliative care team

Refer all undiagnosed chronic cases

**Tertiary level (consult a paediatric gastroenterologist)**

- Stabilise the patient as above
- Management will depend on underlying cause
- Encourage breastfeeding with supplementation with high-caloric-density feeds (involve dieticians)
- Nasogastric tube if oral feeding cannot meet caloric need Fat soluble vitamin supplementation
- (age dependent)
  - Vitamin D (3,000–10,000 IU/day)
  - Vitamin K (2.5–10 mg/day)
  - Vitamin E (10–20mg/kg/day)
  - Vitamin A (5,000–10,000 IU/day)
- Restrict protein intake in children with end-stage liver disease (approximately 2 - 3 g/kg/day)
- Pruritus (multidrug therapy is often required)
  - Cholestyramine (1 - 4 g daily)
  - Phenobarbitone (5 - 10 mg/kg/day)
- Emesis
  - Ondansetron 2 - 4 mg twice daily (<12 years) or 4 - 8 mg twice daily (12 - 18 years). Side effects include worsening of liver function tests
- Coagulopathy
  - Vitamin K provision and use of FFP, cryoprecipitate and platelets as required

- Manage fluid balance and circulatory change (including ascites)
  - Spironolactone (1-9 mg/kg/day in two to three divided doses) and furosemide (0.5-2 mg/kg/day)
  - Salt and water restriction (70% - 80% of maintenance)

**Note:** Avoid vigorous diuretic administration or therapeutic paracentesis as these can further decrease the circulating plasma volume, thereby reducing renal perfusion and increasing sodium retention

- Electrolyte changes and renal failure
  - Correct hypoglycaemia, hypo/hyperkalaemia, hyponatremia, hypocalcaemia and hypomagnesaemia
- Family and psychologic support
  - Physiotherapy may improve gross motor development
  - Ongoing family education
- Palliative care consultation

#### Follow up

- Check for progress of disease/complications

## **Ascites**

### **Definition**

Ascites is the pathologic accumulation of fluid within the peritoneal cavity.

### **Risk factors/causes**

Noncirrhotic ascites:

- Peritoneal inflammation (infections, malignancies, chemical peritonitis [pancreatic and bile- induced ascites])
- Portal venous or lymphatic obstruction (chylous ascites)
- Rupture of intra-abdominal viscus
- Renal (nephrotic syndrome, renal failure)
- Cardiac (pericardial disease, heart failure)
- Hepatic (portal hypertension)

### **Cirrhotic ascites:**

$\alpha$ 1-antitrypsin deficiency, biliary atresia, congenital hepatic fibrosis, neonatal or viral hepatitis, inborn errors of metabolism, storage diseases, autoimmune hepatitis

### **Prevention/promotion**

- Vaccination
- Reduction of alcohol intake
- Early treatment of schistosomiasis

### **Signs and symptoms**

- Respiratory distress present in severe cases
- Protuberant abdomen, bulging flanks, or dullness to percussion in the flanks and everted umbilicus
- Scrotal oedema

### **Investigations and diagnostic testing**

- HIV test
- FBC
- LFTs
- Electrolytes, urea & creatinine
- CRP, ESR
- AFP and LDH
- Serum amylase and lipase

- Blood and urine culture
- Screen for TB, schistosomiasis and syphilis
- Viral serologies (hepatitis A, B, and C, cytomegalovirus and Epstein–Barr virus)
- Diagnostic paracentesis (10–20ml)
- Abdominal ultrasonography
- Abdominal CT or MRI is useful in suspected malignancies

### Ascites fluid analysis studies

Condition	Appearance	Protein**	WBCs/ $\mu$ L	Other tests
Cirrhosis	Straw-coloured or bile-stained	<25 (95%)	<250	
Heart Failure	Straw- coloured	Variable, 15–53	1000	
Nephrosis	Straw- coloured or chylous	<25 (100%)	<250	Sudan staining if chylous
Neoplasm	Straw-coloured, haemorrhagic, or chylous	>25 (75%)	>1000	Cytology, cell block, peritoneal biopsy
Pancreatic ascites	Turbid, chylous, or haemorrhagic	Variable, Often >25	variable	Increased amylase in fluid and serum
Biliary ascites	Bile-stained	Variable, <25	Variable	Increased bilirubin in fluid
TB peritonitis	Clear, turbid, haemorrhagic, or chylous	>25 (50%)	>1000	Stain and culture for acid- fast bacilli, peritoneal biopsy
Pyogenic peritonitis	Turbid or purulent	If purulent, >2.5	>250*	Positive gram stain, culture

\*Normal ascitic fluid contains less than 500 leukocytes/ $\mu$ L with less than 250 polymorphonuclear neutrophils (PMN)/  $\mu$ L. An elevated PMN count greater than 250 cells/ $\mu$ L is a predictor of spontaneous bacterial peritonitis in the absence of bowel perforation.

\*\*Measures in grammes/dL



**Differential diagnosis**

- Mesenteric cysts
- Omental cysts
- Intestinal duplications
- Fluid-filled intestinal loops
- Large ovarian cysts
- Urinary ascites

**Management****Primary level**

Stabilise and refer all cases

**Secondary level**

- Investigate for cause of ascites and manage appropriately
- If no cause established/complicated diagnosis refer to tertiary level for investigations and work as described above

**Tertiary level**

- Stabilise
- Salt restriction
- Restrict fluids when the serum sodium level decreases below 125 mEq/L
  - If fluid resuscitation is indicated, enteral fluids are preferred.
  - **Do not give hypertonic intravenous fluids** as these will increase total body sodium and lead to worsening ascites and fluid overload

**Diuretics**

- Spironolactone
  - 0.5 - 1 mg/kg in infants and 1 - 3 mg/kg in older children (up - 100 mg). It can be increased by 2 mg/kg every 5 - 7 days up to a maximum of 4 - 6 mg/kg/day
- Frusemide
  - Added if there is not an adequate response to spironolactone
  - 1 mg/kg (up to 40 mg). Increase dose by 1 mg/kg to a maximum dose of 4 mg/kg until a response is seen
  - Response to diuresis can be assessed by trending daily weights
- Therapeutic paracentesis
  - Is used in patients unresponsive to medical therapy or if in respiratory distress
  - Can be used periodically to treat refractory ascites
- **Consult gastroenterologist in refractory cases**

**Follow up**

- Review in PEN-Plus clinic every 3 months
- May need routine ascitic taps if chronic ascites

## References

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