Hepatobilliary & Pancreatic diseases

Liver abscess

Diagnostic tools

1. Patient presents with fever usually high grade, comes with chills & rigor; right upper abdominal pain.

2. On examination-patient is febrile, liver is enlarged and tender.

3. Investigation-CBC-neutrophilic leucocytosis, USG of whole abdomen-SOL in liver, may also reveal source of liver abscess like appendicular abscess, biliary obstructive jaundice etc. Blood C/S should be given and if liver abscess drainage done then pus should be sent for C/S.

Organisms causing liver abscess

1. E. histolytica

2. E. coli

3. Pseudomonas

4. Streptococus

5. Staphylococcus etc.

Management

1. Diet- normal

2. Antibiotics before C/S - ampicillin, gentamycin & metronidazole (commonly ciprofloxacin and metronidazole is given).

3. If severe pain then -tramadol HCl.

4. If fever tepid sponging, paracetamol.

5. The underlying source of infection (e.g. biliary disease, dental infection) should be identified and treated.

* Antibiotics are administered for 2-3 weeks, and sometimes up to 6 weeks, metronidazole 800 mg 8 hourly for 7 days.

* Fungal abscess are associated with mortality rates of up to 50% and are treated with intravenous amphotericin B (total dose of 2-9 g) and drainage.

Indication of drainage of liver abscess

1. If the abscess is at least 5 cm in diameter.

2. If the response to antibiotic therapy is poor.

3. Fungal abscess.

4. Other suggested indications for abscess drainage are

-Age of at least 55 years.

-Symptom duration of at least 7 days and

- Involvement of both lobes of the liver.

Acute hepatitis

Diagnostic tools

1. Patient presents with yellow colouration of eye & urine; upper abdominal pain; history of fever, anorexia, nausea, vomiting, joint pain. (nausea and vomiting may be more marked than other symptoms)

2. On examination -jaundice and tender hepatomegaly present.

3. Investigation-s. bilirubin- increase, SGPT-increase, USG of whole abdomen-no billiary obstruction. For cause of acute hepatitis drug history (particularly antitubercular drug, MTX, statins), HBsAg, Anti HAV, anti HEV should be done.

Management

1. Complete bed rest.

2. Diet- normal

3. Infusion 5% DNS 1000 cc i/v @ 20 drops/ minute stat & daily (If nausea and vomiting are pronounced or if oral intake is substantially decreased).

4. Tab. Ursocol (300mg) 1+0+1 (if feature of cholestasis e.g. itching, steatorrhoea).

5. No paracetamol/ NSAIDs/ sedatives, antiviral, IM injection.

6. Monitor for liver failure e.g. confusion, disorientation, loss of consciousness, altered sleep pattern, any bleeding manifestation etc.

7. Patient should be return to normal activities when serum bilirubin, SGPT is normal and patient have no symptoms.

Follow up checklist

1. Symptomatic improvement or not.

2. Appearance of new sign, symptoms e.g. itching, steatorrhoea etc.

3. Orientation, sleep pattern normal or not.

4. Bowel and bladder movement normal or not.

5. Pulse, blood pressure.

6. Flapping tremor.

Indication of hospitalization of hepatitis patient

a) Features of hepatic encephalopathy.

b) Pronounced anorexia, vomiting requiring parental nutrition.

c) S. bilirubin >15 mg/dl.

d) Prolonged prothrombin time.

e) Co-morbidities- like DM, IHD, CKD.

Criteria of hospital discharge

a) Symptomatic improvement.

b) A significant downward trend of SGPT and serum bilirubin level.

c) Normal prothrombin time.

d) Mild aminotransferase elevations should not be considered contraindications to the gradual resumption of normal activity.

Acute liver failure

Hyperacute liver failure is if appearance of jaundice to encephalopathy develops time within a period of less than 7 days. Occurs in viral hepatitis and paracetamol toxicity, cerebral edema is common.

Acute liver failure is if appearance of jaundice to encephalopathy develops time within a period of 7-28 days. Cerebral edema is common. Usually cryptogenic or drug induced.

Subacute liver failure is if appearance of jaundice to encephalopathy develops time 29 days-12 weeks.

Diagnostic tools

1. Yellow coloration of eye & urine or diagnosed case of acute hepatitis.

2. Alteration of consciousness.

3. On examination-jaundice present

- Patient may be drowsy, disoriented or unconscious.

-Flapping tremor may be present.

-Planter bilateral extensor.

4. Investigation-diagnosis is clinical; to determine cause of liver failure history of fever, drug history (taking paracetamol, NSAID, diuretic) should be taken, serum electrolytes, viral markers HBsAg, anti HAV, anti HEV should be checked to determine the cause of liver failure.

Management-definitive management is liver transplantation & N-acetylcystein in paracetamol induced liver failure.

Conservative management (should be in ICU)

1. NG tube feeding if patient is unconscious.

2. Infusion 10% DA 1000 cc iv @ 10 drops/minute stat. & daily.

3. Broad spectrum antibiotic (ceftriaxone and metronidazole).

4. Omeprazole sachet 1 sachet dissolve in ½ glass water then take via NG tube 12 hourly.

5. Syrp. Lactulose 2 TSF TDS (if constipation present; dose can be increase up to 6 TSF 8 hourly).

6. Vitamin K 10 mg daily for 3 days.

7. Syrp. UDCA 2TSF TDS.

8. Continued catheterization.

9. Change the posture 2 hourly.

10. Monitor blood glucose (as hypoglycaemia may occur) vital signs.

Follow up checklist

1. Conscious level by GCS.

2. Bowel movement.

3. Flapping tremor.

4. Pulse, BP, urine output.

N:B: Target of bowel movement is 2 times daily, if not achieved then increased dose of lactulose from 2TSF to 6 TSF 3 times daily. If still target not achieved then add enema simplex.

Chronic liver disease

Diagnostic tools

CLD patient usually present with complications like ascites, haematemesis & melaena, encephalopathy etc. other mode of presentations are-

1. Patient present with swelling of abdomen.

2. History of jaundice in past (may be absent, because infection may be asmptomatic).

3. On examination-ascitis & splenomegaly.

4. Investigation

    a) LFT-low serum albumin, altered albumin : globulin ratio.

    b) Ascites fluid study-transudative ascites.

    c) USG feature suggestive of CLD & splenomegaly.

    d) Endoscopy of upper GIT may reveal esophageal varices.

Management

1. General Measures

i. Abstinence from alcohol (If patient is alcoholic).

ii. Fluid (1-1.5 L/day) & salt restriction (no added salt).

iii. Patient with cirrhosis should receive the HAV, HBV, and pneumococcal vaccines and a yearly influenza vaccine.

iv. Complete bed rest.

2. Diuretics- spironolactone alone or combine with frusemide (usually starting dose is spironolactone 100 mg, with doubling the dose after 48-72 hours if no desirable diuresis, after achieving highest dose 400 mg daily, frusemide can be added). (Diuretics should be titrated to remove no more than 1 L of fluid daily)

3. Primary prevention of variceal bleeding

a) Propranolol (80–160 mg/day) or nadolol (40–240mg/day). (dose should be titrated to reduce heart rate 25%). Or

b) Carvedilol is also effective at doses of 6.25–12.5 mg/day, (dose should be titrated to achieve a heart rate of 50–55 beats/min, if possible). Or

c) Prophylactic banding is also effective and should be considered for those unable to tolerate drugs.

4. If cause of CLD is viral then add antiviral

5. If there is constipation then syrp. lactulose 2 TSF TDS (can be increased upto 6 TSF TDS)

6. Regular follow up the patient with pulse, BP, flapping tremor, weight, abdominal girth.

Paracentesis

Paracentesis to dryness is safe, provided the circulation is supported with an intravenous colloid such as human albumin (6–8 g per litre of ascites removed, usually as 100 mL of 20% or 25% human albumin solution (HAS) for every 1.5–2 L of ascites drained) or another plasma expander. Paracentesis can be used as an initial therapy.

Other indications of paracentesis of ascites fluid are

a) In patients with massive ascites and respiratory compromise.

b) Ascites refractory to diuretics or

c) Intolerable diuretic side effects of diuretic

N:B:

a) In patients who cannot tolerate spironolactone because of side effects, such as painful gynecomastia, amiloride (another potassium-sparing diuretic) may be used in a dose of 5-10 mg orally daily.

b) Target of bowel movement is 2 times daily. Lactulose can be used up to 6 TSF 8 hourly.

c) Diuretics should be titrated to remove no more than 1 L of fluid daily, so body weight should not fall by more than 1 kg daily to avoid excessive fluid depletion.

Haematemesis & Melaena

Diagnostic tool

1. History of vomiting out of blood and passes of black tarry stool. There may be history of taking NSAIDs, PUD, CLD

2. On examination - pulse tachycardia, BP- may be hypotensive, epigastric tenderness/mass, feature of CLD - splenomegaly, ascites may be present.

3. Investigation-endoscopy of upper GIT may reveal ulcer of PUD, rupture oesophageal varices, carcinoma of stomach etc.

Management

1. NPO TFO

2. Assessment -it is mandatory to assess the patient (by pulse, blood pressure, respiration, urine output), whether shock present or not and severity of bleeding. Usually haematemesis is severe.

3. Infusion normal saline/Hartman saline if patient is hypotensive or in shock.

4. Infusion 5% DA 1000 cc + 5% DNS 1000 cc IV @ 20 drops/minute stat and daily.

5. Inj. Esomeprazole (40mg) 2 vial IV stat slowly over 10 minutes then

5% DA 500 cc + Inj. Esomeprazole (40mg) 3 vial IV @ 8 drops /minute.

Then

Infusion 5% DA 500 cc + Inj. Esomeprazole (40mg) 2 vials) IV @ 8 drops /minute daily.

(Commonly injection esomeprazole 40 mg 2 vial IV stat over 10 minutes then 1 vial IV over 10 minutes 12 hourly is given).

6. In diagnosed CLD patient-prophylactic broadspectrum antibiotics, such as oral ciprofloxacin or intravenous cephalosporin or piperacillin/tazobactam.

7. Blood transfusion- indication

- If patient is in shock

-or rebleeding/ massive bleeding occurs.

(Feature of rebleeding are fresh haematemesis & melaena with shock or fall of Hb >2gm/dl in 24 hours).

8. Close monitoring of the patient with pulse, BP, urine output and Hb%.

9. Endoscopy-should be carried out after adequate resuscitation, ideally within 24 hours.

a) Patients who are found to have major endoscopic stigmata of recent haemorrhage can be treated endoscopically using a thermal or mechanical modality, such as a ‘heater probe’or endoscopic clips, combined with injection of dilute adrenaline (epinephrine) into the bleeding point (‘dual therapy’). A biologically inert haemostatic mineral powder (TC325, ‘haemospray’) can be used as rescue therapy when standard therapy fails.

b) If bleeding from varices then vasoactive medications (e.g. terlipressin-2 mg IV 4 times daily until bleeding stops, and then 1 mg 4 times daily for up to 72 hours.), endoscopic therapy (banding or sclerotherapy), balloon tamponade, TIPSS and, rarely oesophageal transection can be done.

10. Oral food can be allowed if there is no haematemesis and melaena for last 48 hours.

Tips

* A systolic blood pressure less than 100 mm Hg identifies a high risk patient with severe acute bleeding.

* A heart rate over 100 beats/minute with a systolic blood pressure over 100 mm Hg signifies moderate acute blood loss.

* A normal systolic blood pressure and heart rate suggest relatively minor hemorrhage.

* In actively bleeding patients, platelets are transfused if the platelet count is under 50,000/mm^3 and considered if there is impaired platelet function due to aspirin or clopidogrel use (regardless of the platelet count).

* Uremic patients (who also have dysfunctional platelets) with active bleeding are given three doses of desmopressin (DDAVP), 0.3 mcg/kg intravenously, at 12-hour intervals.

* Fresh frozen plasma is administered for actively bleeding patients with a coagulopathy and an INR > 1.5.

* If massive bleeding, 1 unit of fresh frozen plasma should be given for each 5 units of packed red blood cells transfused.

* Predictor of rebleeding- clinical predictors of increased risk of rebleeding and death include

a. Age > 60 years

b. Co morbid illnesses

c. Systolic blood pressure < 100 mm Hg

d. Pulse > 100 beats/minute and

e. Bright red blood in the nasogastric aspirate or on rectal examination.

Features of active bleeding: Patients with active bleeding manifested by hematemesis or bright red blood on nasogastric aspirate, shock, persistent hemodynamic derangement despite fluid resuscitation.

Acute pancreatitis

Diagnostic tool

1. Patient presents with-severe constant upper abdominal pain may radiate to back, relieve by sitting and leaning forward, fever & vomiting.

2. On examination -upper abdominal tenderness but no rigidity and rebound tenderness.

3. Investigation- serum amylase increase, USG of whole abdomen may reveal swelling of the pancreas.

Urgent Investigation

1. S. amylase (if patient present within 48 hours of onset of abdominal pain).

2. S. lipase (if patient present after 48 hours of onset of abdominal pain).

3. ECG (to exclude MI).

4. USG of whole abdomen.

Management

1. NPO TFO (oral intake of fluid and foods can be resumed when the patient is largely free of pain and has bowel sounds (even if the serum amylase is still elevated).

2. Nasogastric aspiration is required only if paralytic ileus is present.

3. O2 inhalation 4-6 L/ minute SOS.

4. Infusion 5% DA 1000 cc + 5% DNS 1000 cc IV @ 20 drops/ minute stat and daily.

5. Antibiotic-carbapenems or quinolones and metronidazole.

6. Inj. Omeprazole (40mg) 1 vial IV stat over 10 minutes & daily.

7. Analgesic-opoid analgesic (nalbuphine, morphine, pethidine) should be used. Usually tramadol HCl either injection or suppository form use in ward.

8. Prophylaxis of thromboembolism with subcutaneous low molecular weight heparin.

9. Close monitoring of the patient with pulse, BP and urine output.

N: B:

* If cause of acute pancreatitis is gall stone then surgery should be performed after 2 weeks of recovery.

Chronic pancreatitis

Diagnostic tools

1. Patient present with recurrent upper abdominal pain, vomiting, diarrhoea, features of DM. Patient may present with features of acute pancreatitis (acute on chronic pancreatitis).

2. On examination-patient is lean thin (features of malnutrition), upper abdominal tenderness may be present.

3. Investigation- USG of whole abdomen may reveal pancreatic calculi, X-ray of abdomen may reveal pancreatic calcification.

Management

1. Treatment of the underlying cause (avoidance of alcohol, pancreatic stone removal etc).

2. Analgesic

    a) NSAID or opiate.

    b) Pregabalin and tricyclic antidepressants at a low dose, may be effective.

    c) Coeliac plexus neurolysis produces long lasting pain relief.

3. For malabsorption

a) Dietary fat restriction (with supplementary medium chain triglyceride therapy in malnourished patients).

b) Oral pancreatic enzyme supplement-pancreatin 1-3 tablets daily.

c) PPI

4. Management of DM-insulin.

5. Patients who are abstinent from alcohol and who have severe chronic pain that is resistant to conservative measures should be considered for surgical or endoscopic pancreatic therapy.

6. Management of complications-surgical or endoscopic therapy may be necessary for the management of pseudocysts, pancreatic ascites, common bile duct or duodenal stricture and the consequences of portal hypertension.

Variceal bleeding

Diagnostic tools

1. This patient usually present with haematemesis and melaena, patient may be diagnosed case of CLD. Features of CLD may be predominant symptoms.

2. Endoscopy of upper GIT-usually reveal rupture esophageal varices.

Management

1. Same as management of haematemesis and melaena.

2. Prophylactic antibiotic-prophylactic broadspectrum antibiotics, such as oral ciprofloxacin or intravenous cephalosporin or piperacillin/tazobactam.

3. Vasopressor (terlipressin)-2 mg IV 6 hourly until bleeding stops then 1 mg 6 hourly for up to 72 hours.

4. Endoscopic procedure to stop variceal bleeding-

* Band ligation-should be repeated 2-4 week until varices are obliterated. Regular follow up endoscopy is required to identify & treatment of any recurrence of varice.

* Ballon tamponade- Sengstaken-Blakemore tube to stop active bleeding.

5. TIPSS- patient in whom other treatment is not successful & those with good liver function.

6. Oesophageal transection

- When TIPSS is not available

- Bleeding cannot be controlled with other therapies.

Portal hypertension

Main complication of portal hypertension is haematemesis and melaena.

Diagnostic tools

1. Patient may be diagnosed case of CLD.

2. Features of portal hypertension are-engorged vein in upper abdomen, ascites, splenomegaly and present of oesophageal varices in endoscopic examination.

Management

1. Primary prevention of variceal bleeding

a) Propranolol (80–160 mg/day) or nadolol (40–240 mg/day). (dose should be titrated to reduce heart rate 25%). Or

b) Carvedilol is also effective at doses of 6.25–12.5 mg/day, (dose should be titrated to achieve a heart rate of 50–55 beats/min, if possible).

Or

c) Prophylactic banding is also effective and should be considered who unable to tolerate drugs.

2. Secondary prevention of variceal bleeding (to prevent variceal bleeding in patient who already have variceal bleeding)-beta blocker following endoscopic variceal banding.

Wilson's disease

Diagnostic tools

1. Features of CLD, patient usually young, may complaint recurrent jaundice and features of parkinsonism and psychological problems may be present.

2. On examination-stigmata of CLD and K-F ring may be present.

3. Investigation-same as CLD, serum caeruloplasmin usually low. Serum copper-high. Measuring 24-hour urinary copper excretion while giving D-penicillamine is a useful confirmatory test; more than 25 μmol/24 hrs is considered diagnostic of Wilson's disease.

Management

1. Penicillamine -most patient require 1.5 gm/day (1-4gm/day). The dose can be reduced once the disease is in remission. Treatment should be continued throughout the life including pregnancy. Abrupt discontinuation may precipitate acute liver failure

2. Trientrine dihydrochloride 1.2-2.4 gm/day & Zinc 50 mg 8 hourly, if side effects of penicillamine occur.

3. Liver transplantation

- In fulminant liver failure

- Advance cirrhosis with liver failure.

4. Siblings & children of patient with Wilson's disease must be investigated & treatment should be given to all affected individual even if they are asymptomatic.

Autoimmune Hepatitis

Diagnostic tools

1. Features of acute hepatitis, but resolution does not occur, fever, arthralgia, vitiligo and epistaxis may be present. Features of other autoimmune disease (Hashimoto's thyroiditis or rheumatoid arthritis) may be present.

2. Investigation-ANA, anti smooth muscle antibody positive. Liver biopsy confirm the diagnosis.

Management

Prednisolone 40 mg daily

-Dose can be reduced as the patient & LFT improve. Maintainance therapy (prednisolone below 5-10 mg/day) is required after LFT have returned to normal & withdrawl of treatment should not be considered unless liver biopsy is also normal.

-Azathioprine 1-1.5 mg/kg/day allows the dose of prednisolone to be reduced.

Primary billiary cholangitis (previously known as primary billiary cirrhosis)

Diagnostic tools

1. Female patient, middle age, history of flactuating jaundice and itching. On examination- xanthoma may be found, mild hepatomegaly is common and splenomegaly becomes increasingly common as portal hypertension develops.

2. Investigation- the LFTs shows a pattern of cholestasis, hypercholesterolaemia is common. antimitochondrial antibody positive; USG and ERCP to exclude other billiary disease.

Management

1. 1st line drug-UDCA 13-15 mg/kg/day.

2. 2nd line drug-Obeticholic acid (OCA), for patients showing an inadequate response to UDCA.

2. Liver transplantation if

- Liver failure

- Intractable pruritus.

3. Pruritus (occurs due to up regulation of opoid receptor & increase endogenous opoid). Managed by

i) Cholestyramine 4-16gm/day, the powder mixed in orange juice then taken before & after breakfast. It is ineffective in complete billiary obstruction.

ii) Alternative treatment-rifampicin 150 mg/day (highest dose 600 mg/day), naltrexone (opoid antagonist) 25 mg/ day up to 300 mg /day, plasmapheresis and liver support device (e.g. a molecular adsorbent recirculating system, MARS).

4.Fatigue-no treatmnent, exclude depression & hypothyroidism.

5. Supplementation of fat soluble vitamins

6. Bone disease

Replacement of calcium & vit-D3.

Bisphosphonate if evidence of osteoporesis.

Acute cholecystitis

Diagnostic tools

1. Patient presents with severe upper abdominal pain (right upper quadrant) with vomiting, may have history of fever.

2. On examination-Mc Burney's point tender.

3. Investigation-CBC-leucocytosis, ultrasonography detects gallstones and gallbladder wall thickening.

Management

1. NPO TFO.

2. IV Fluid- infusion 5% DA 1000 cc + 5% DNS 1000 cc i/v @ 20 drops/ minute stat and daily.

3. Intravenous omeprazole 40 mg 1 vial IV over 10 minutes stat and daily.

4. Analgesic-moderate pain-NSAID, severe pain-pethidine/ pentazocin.

5. Antibiotic-cephalosporin (cefuroxime), if severely ill add metronidazole.

6. NG aspiration- if persisting vomiting.

7. Urgent surgery-when cholecystitis progresses in spite of medical therapy and when complications such as empyema or perforation develop.

Alcoholic liver disease (ALD)

In the UK, a unit of alcohol contains 8 g of ethanol. An upper threshold of 14 units/week in women and 21 units/week in men is generally considered safe. The risk threshold for developing ALD is variable but begins at 30 g/day (3.75 units/day) of ethanol. Clinical features of ALD

1. ALD has a wide clinical spectrum, ranging from mild abnormalities of LFTs on biochemical testing to advanced cirrhosis.

2. The liver is often enlarged in ALD, even in the presence of cirrhosis.

3. Stigmata of chronic liver disease, such as palmar erythema, are more common in alcoholic cirrhosis than in cirrhosis of other aetiologies.

Features of alcoholic fatty liver disease

1. Elevated transaminases and absence of hepatomegaly.

2. It has a good prognosis and steatosis usually disappears after 3 months of abstinence.

Features of alcoholic hepatitis

1. Patient has jaundice and hepatomegaly

2. Complications of portal hypertension may also be present.

3. In patients presenting with jaundice who subsequently abstain, the 3- and 5-year survival is 70%. In contrast, those who continue to drink have 3- and 5-year survival rates of 60% and 34%, respectively.

Features of alcoholic cirrhosis

1. Alcoholic cirrhosis often presents with a serious complication, such as variceal haemorrhage or ascites.

2. Only half of such patients will survive for 5 years from presentation. However, most who survive the initial illness and who become abstinent will survive beyond 5 years.

Investigation

1. Support of alcohol intake- macrocytosis in the absence of anaemia, may suggest and support a history of alcohol misuse.

2. LFT

3. Investigation to detect other alcohol related diseases

Management

1. Cessation of alcohol

2. Nutrition should be maintained

3. Steroid (prednisolone)

Indication- Severe alcoholic hepatitis (Maddreys discrimination score >32) (calculation of Maddreys DF = [4.6 × Increase in PT (sec)] +Bilirubin (mg/dL).

Contraindication:

i) Existing sepsis

ii) Variceal haemorrhage

(If bilirubin has not fallen 7 days after starting therapy in treatment with steroid, then steroid is unlikely to reduce mortality & should be stopped).

4. Pentoxifylline-use in severe alcoholic hepatitis, it appears to reduce incidence of hepato-renal failure, its use is not complicated by sepsis.

5. Liver transplantation

NAFLD

It can be classified into fatty infiltration alone (steatosis) to fatty infiltration with inflammation (non-alcoholic steatohepatitis, NASH) and may progress to cirrhosis and primary liver cancer.

Diagnostic tools

1. NAFLD is frequently asymptomatic, although it may be associated with fatigue and mild right upper quadrant discomfort.

2. Usually diagnosed incidentally during routine biochemical test or as a fatty liver during an ultrasound or CT scan of the abdomen.

3. May present with complications of cirrhosis and portal hypertension, such as variceal haemorrhage, or with hepatocellular carcinoma.

4. Investigation

a) Exclusion of excess alcohol consumption and other liver diseases (including viral, autoimmune and other metabolic causes).

b) ALT & AST. NASH- AST: ALT ratio of < 1, Cirrhosis- AST: ALT ratio of > 1.

c) Imaging- ultrasound is most often used and provides a qualitative assessment of hepatic fat content, CT, MRI or MR spectroscopy offer greater sensitivity for detecting lesser degrees of steatosis.

d) Liver biopsy remains the ‘gold standard’ investigation for diagnosis and assessment of degree of inflammation and extent of liver fibrosis.

Management

1. NAFLD requires no treatment except-reduction of body weight.

2. Statin to treat dyslipidaemia.

3. Specific insulin-sensitising agents, in particular glitazones, may help selected patients, while recent results with bezafibrate, a lipid-lowering fibrate, have been encouraging.

Haemochromatosis

Haemochromatosis may be primary and secondary. Commonly we face secondary haemochromatosis occurs due to haemolytic anaemia and other condition requiring multiple blood transfusion.

Diagnostic tools

1. Usually patient men and over 40 years

2. Usually present with features of liver disease (often with hepatomegaly), type 2 diabetes or heart failure.

3. Leaden-grey skin pigmentation due to excess melanin occurs, especially in exposed parts, axillae, groins and genitalia: hence the term ‘bronzed diabetes’.

4. Impotence, loss of libido and testicular atrophy are recognized complications

5. Investigation

a) Iron profiles- increased ferritin, a raised plasma iron and saturated plasma iron-binding capacity.

b) Liver biopsy allows assessment of fibrosis and distribu tion of iron (hepatocyte iron characteristic of haemochro matosis).

c) The Hepatic Iron Index (HII) provides quantification of liver iron (μmol of iron per g dry weight of liver/age in years). An HII of more than 1.9 suggests genetic haemochromatosis.

Management

Primary haemochromatosis

1. Weekly venesection of 500 ml of blood (250 mg of iron) until s. iron is normal; this may take up to 2 years or more. The aim is to reduce ferittin <50 µg/ L.

2. Treatment of associated DM & cirrhosis (if any).

3. Asymptomatic 1st degree family members should be investigated, preferably by genetic screening & plasma feritin & iron binding saturation. Liver biopsy is indicated if

-LFT abnormal

-And/or S. feritin is>1000 µg/ L.

4. Asymptomatic disease also should be treated by venesec tion.

5. Screening of HCC by USG.

Secondary haemochromatosis

1. Treatment of the underlying cause and desfuroxamine (iron chelating agent)