Abdominal PACES

1. Splenomegaly
2. Patient on dialysis
3. Polycystic kidney disease
4. Renal transplant
5. Chronic liver disease
6. Hepatosplenomegaly
7. Ascites
8. Unilateral enlarged kidney
9. Crohn’s disease
10. PBC
11. Liver transplant
12. Jaundice
Chronic liver disease

History:
Fatigue, weight loss, jaundice, increasing abdominal girth, mental state changes

Cause:
- Alcohol
- Hep B of C – transfusion, sex, IVDU
- Meds – Amiodarone, methylldopa, methotrexate
- Diabetes? Tan? Haemochromatosis
- Chorea? Wilsons
- Emphysema? Alpha1AT
- FHx of probs

Signs:
- Clubbing
- Leukonychia
- Dupreylten’s contracture
- Palmar erythema
- Spider naevi
- Tattoos
- Hepatic flap
- Pallor
- Scratch marks
- Pigmentation
- Icterus
- Parotid enlargement
- Loss of hair
- Gynaecomastia
- ?splenomegaly and ascites
- ?hepatomegaly

Causes:
- Alcohol is the most common in the UK
- Viral hepatitis B and C is next most common, and the most common worldwide
- Autoimmune – PBC, PSC, AIH 1 and 2
- Metabolic – Haemochromatosis, Wilson’s, alpha1AT
- Drugs – Methotrexate, Amiodarone
- Cryptogenic

Investigations
- Routine bloods and liver enzymes and function
- Blood tests for causes
- AFP
Complications:
  • From hepatic dysfunction:
    o Encephalopathy
    o Coagulopathy
    o HCC
    o Nutritional problems
  • From Portal hypertension:
    o Ascites
    o SBP
    o Variceal bleeding
    o Hepatorenal syndrome

Child Pugh – Prognosis from chronic liver Dx (Synthetic and metabolic function)
  • Bilirubin
  • Albumin
  • INR
  • Ascites
  • Encephalopathy

MELD score – equivalent
  • Bilirubin
  • INR
  • Cr
Primary Biliary cirrhosis

History:
- Fatigue
- Pruritis
- Jaundice
- Xanthomata
- CLD complications

Examination:
- Middle aged woman
- Icteric with skin pigmentation
- Xanthelasma/xanthomata
- Excoriations
- Hepato/hepatosplenomegaly

What is PBC?
An autoimmune disease of the liver with progressive destructions of small bile canaliculi leading to cholestasis, fibrosis and cirrhosis. 90% female. It is associated with a range of other autoimmune conditions including sjögren’s, RA, SS, Hashimotos, Coeliac’s. It may be triggered by exposure to a pathogen.

How is it diagnosed?
- Autoantibodies – AMA (95%), SMA (50%), ANA (20%)
- Abdo US and elastography
- Liver biopsy to stage disease in some patients

How is it treated?
- Ursodeoxycholic acid – improved cholestasis and blood tests. Little effect on symptoms. It may help progression. It reduces cholesterol absorption.
- Cholestyramine (bile acid sequestrant) reduces itching
- Avoidance of hepatotoxins, esp alcohol
- Dietitian input with vitamin supplementation (esp fat soluble ones)
- Liver transplantation (but 30% recur in 10 years)
**Idiopathic Haemochromatosis**

**Examination:**
- Slate grey pigmentation
- Decreased body hair
- Gynaecomastia
- Testicular atrophy
- Hepatosplenomegaly with CLD

**Other signs:**
- Cardiomyopathy +/- arrhythmias
- Diabetes
- Arthropathy
- Addison’s disease
- Hypothyroidism
- Hypogonadism

**What is it?**
Hereditary haemochromatosis is a genetic, autosomal recessive, iron storage disorder characterised by a state of iron overload, which causes damage to a range of organs. Classical (type 1) HH caused by HFE gene mutation, however other forms involving hepcidin, transferrin and ferroportin mutations are known. Secondary haemochromatosis can occur from chronic haemolysis, repeated blood transfusion or excess iron intake (rarely).

**Investigations:**
- Haematinics – ferreting and transferring saturations.
- HFE genetic analysis
- MRI
- Liver Bx

**Treatment:**
- Venesection – aim for ferritin below a certain level.
- Desferoxamine – chelation agent – infusion (P in the A)
- Exjade – chelation agent – oral
- Genetic counselling and family testing
Splenomegaly

Massive splenomegaly:

- Myeloproliferative disorders:
  - CML
  - Myelofibrosis
  - PRV
- Storage diseases
  - Gauchers
- Tropical infection
  - Chronic malaria
  - Visceral leishmaniasis
- Haematological malignancy
  - Lymphomas and Leukaemias

Moderate splenomegaly

- Myeloproliferative disorders:
  - CML
  - Myelofibrosis
  - PRV
- Haematological malignancy
  - Lymphomas and Leukaemias
- Cirrhosis with portal hypertension

Mild splenomegaly

- Myeloproliferative disorders:
  - CML
  - Myelofibrosis
  - PRV
- Haematological malignancy
  - Lymphomas and Leukaemias
- Cirrhosis with portal hypertension
- Infections:
  - EBV, CMV
  - Endocarditis
  - Hepatitis
  - HIV
  - Brucellosis
  - Toxoplasmosis
- Haemolysis
Indications for splenectomy:

- Haemorrhage – traumatic or atraumatic
- Significant transfusion requirements in haemolytic disorders such as Thalassaemia
- Significant symptoms from splenic enlargement
- Leucopenia or thrombocytopenia due to hypersplenism

Complications of splenectomy:

- Thrombocytosis
- Overwhelming sepsis from encapsulated organisms.
- Prevent with vaccination (Pneumococcal, HiB, Meningitis, Flu) and prophylaxis
Hepatomegaly

Examination:

Size, tenderness, surface, span, bruit
Is there a spleen?
Is there a spleen and ascites?
Are there signs of CLD?
Are their LNs?
Is there an indication of a primary tumour?
Is there a raised JVP/signs of right heart failure?

Top 3
1. Liver fibrosis/chronic hepatitis/cirrhosis – most likely alcohol (signs of CLD)
2. Mitotic lesion – (evidence of primary, clubbing, cachexia, irregular)
3. CCF – (JVP, S3, pulm heave, ankle oedema, TR, giant V waves, pulsatile liver)

Others:
- **Infections** – Hepatitis, EBV
- **Neoplasms of the liver** – benign or malignant
- **Lymphoproliferative Dx** (LNs)
- **Primary biliary cirrhosis** (middle aged female with jaundice, scratchmarks and xanthelasma)
- **Haemochromatosis** (male, grey pigmentation)
- **Sarcoidosis** (EN, lupus pernio, MZ crackles, skin signs)
- Reidel’s lobe
- Emphysema with apparent hepatomegaly
- **Amyloidosis** (RA or other chronic Dx)
- Budd-Chiari (icterus, ascites, tender hepatomegaly)
- Hydatid cyst (?Welsh?!?)
- Amoebic abscess (drain sites, ethnic origin)

?Hard and nobbly liver?
- **Malignancy**
- Polycystic liver Dx (feel for kidneys)
- Macronodular cirrhosis (rare)
- Hydatid cysts
- Syphilitic gummata
Hepatosplenomegaly

Think about associated symptoms and the size of the organs:

HSM with Lymphadenopathy
- CLL
- Lymphoma
- Infectious mononucleosis
- Infective hepatitis
- Sarcoidosis
- Amyloidosis
- Tropical infection – Brucellosis, CMV, Toxo
- HIV with PGL

With signs of Chronic liver disease
   Cirrhosis with portal hypertension

With jaundice
   Haemolysis (hereditary spherocytosis, sickle cell disease, thalassaemia)

With anaemia
   Myeloproliferative disease

Tell me about Polycythaemia rubra vera?
- Increased RBC mass without secondary causes
- Increased BM activity with high RCC, high WCC, high Plts
- Decreased MCV with iron deficiency
- JAK2 positive

Symptoms and signs
- Facial plethora
- Ecchymoses and easy bruising
- Itching/scratch marks
- Engorged conjunctival vessels
- Dilated retinal veins
- High BP
- Splenomegaly

Secondary polycythaemia?
- Dehydration/Diuretics
- Low Art O2 – COPD, R→L shunt, OSA
- Abnormal O2 release – smokers, carboxyhaemoglobinemia
- Cobalt poisoning
- Inappropriate EPO – renal/adrenal/liver tumours, factitious, APCKD
What is the management for an asplenic patient?
• Vaccination – HiB, Meningococcal, Pneumococcal, Influenza
• Prophylactic Penicillin V
• Education of patient, family, GP of risk of overwhelming infection
• Medic alert bracelet

How do you diagnose and stage Lymphoma?
• LN Biopsy – preferably excision
• CT NCAP
• Hx for B symptoms
• +/-
• PET
• ?BM aspirate
• ?ENT exam
• ?CSF Ax

→ Ann Arbor staging – originally for Hodgkin’s, now for NHL too.
Ascites

Demonstrate shifting dullness  
Look for resp compromise  
Look for SBP  
Look for causes!!!  
- CLD  
- Cancer  
- CCF  
- TB

Causes

Top 5
1. Cirrhosis with portal hypertension (signs of CLD and indications of cause)  
2. Intra-abdominal malignancy (scars, hard knobbly liver, mass, cachexia, Troissiers, LNs)  
3. CCF (signs of right heart failure)  
4. Nephrotic syndrome (puffy eyes, swollen legs, chronic disease causing amyloid or DM)  
5. TB peritonitis (ethnic origin, chest)

Other
- Constrictive pericarditis  
- Budd Chiari syndrome  
- Myxoedema  
- Meig’s syndrome  
- Chylous ascites
Bilateral renal enlargement

Look for:
- Aspiration scars
- AV fistula/haemodialysis
- Peritoneal dialysis catheter
- Transplant
- BP

Causes:
- Adult Polycystic Kidney Disease
- Bilateral hydronephrosis
- Amyloidosis
- Tuberous sclerosis with non-malignant neoplasms
- VHL Dx – tumours – more likely cancerous in origin

Unilateral renal enlargement
- Polycystic kidney Dx (either with only 1 palpable or one removed)
- Mitotic lesion
- Unilateral hydronephrosis
- Hypertrophy of single functioning kidney
Transplanted kidney

Scar in RIF
- Mass underneath scar – dull to percussion, possibly a bruit, non-tender (we hope)

Look for causes:
- Diabetes – Insulin, marks on abdomen, needle marks on fingers, ophthalmoscopy
- Hypertension – ask for BP
- Adult polycystic kidney disease (bilateral renal masses or nephrectomy scars)

Look for previous replacement
- AV fistula – patent/ligated, recently needled
- Old Hickman/Portacath site
- Old Peritoneal dialysis scars

Look for complications:
- Uraemic? Requiring treatment (needled fistula)
- Ciclosporin signs (gum hypertrophy, hypertrichosis, skin lesions)
- Look for infections from immunosuppression
- Look for renal tenderness
- Renal bruit – renal artery stenosis
Kidney questions:

How would you test to see if the graft is working?
- Examination – tender, bruit, uraemic encephalopathy
- Urine volume in 24 hours
- Urinary protein on dip or collection
- Bloods – deteriorating renal function
- Consider biopsy with specialist opinion

What are the complications of renal transplant?
- Graft rejection
- Disease recurrence
  - Haemolytic uraemic syndrome
  - FSGC
  - Cystinosis
  - Fabry’s disease
- Renal artery stenosis
- Opportunistic infections
- Drug SE’s
- IHD
- Hypertension
- Skin cancer and lymphoma

Causes of gum hypertrophy?
- Scurvy
- Leukaemia
- Ciclosporin
- Phenytoin
- Nifedipine

Drugs used for immunosuppression?
Ciclosporin
- Calcineurin inhibition $\rightarrow$ decreased IL2
- Nephrotoxic
- Hepatic dysfunction
- Gum hypertrophy and hypertrichosis
- Hypertension
- High lipids and glucose

Azathioprine
- BM suppression (test for TPMT)
- Hepatotoxic

Corticosteroids

Tacrolimus
- Calcineurin inhibitor
- Diabetogenic (and BM suppression)
Complications of APCKD?
3rd most common cause of CKD in UK. 1:1000 prevalence. Auto Dominant.
Renal
• Loin pain from size or haemorrhage
• Hypertension
• Renal failure
• Anaemia
• Polycythaemia
Extra-renal
• Liver, pancreas, spleen cysts
• Mitral valve prolapse
• Sub-arachnoid haemorrhage
• Colonic diverticulae

Renal replacement therapy – pros and cons
Haemodialysis
• 3x4hour sessions a week only
• Better electrolyte control
• Less infection risk
• BUT:
  o Fistula formation and complications such as thrombosis and aneurysms
  o Portacath or neck line insertion
  o Must travel
Peritoneal dialysis
• Low tech, low cost
• Done at home, when convenient
• No surgery needed
• BUT:
  o Peritoneal catheter must be placed
  o 3 bags a day – time consuming
  o risk of infection

Indications for urgent dialysis:
• Uraemic Pericarditis/Pleuritis
• Encephalopathy/Neuropathy
• Bleeding diathesis
• Resistant fluid overload
• Resistant electrolyte abnormality
• Resistant malignant hypertension
**Contraindications for dialysis:**
- Predicted survival less than 5 years
- Predicted risk of graft loss more than 50% at 1 year
- Unable to comply with immunosuppressant therapy
- Risk of life-threatening complications of immunosuppression (chronic viral illness, malignancy)

**Mesangiocapillary GN:**
Type 1 – Immune deposits e.g. cryoglobulinaemia in Hep C
Type 2 – Familial with lipoatrophy
Type 3 – similar to type 1 without cryoglobulinaemia

**What are the complications of chronic renal failure?**

**Cardiovascular**
- Hypertension – control aggressively
- Volume overload
- Increased IHD
- Decreased Hb due to EPO decreasing – give EPO

**Metabolic**
- Uremia
- Potassium rise
- Metabolic acidosis

**Bone**
- Hyperphosphataemia
- Hypocalcaemia due to lack of hydroxylation of vitamin D
- Hypercalcaemia due to secondary then tertiary hyperparathyroidism
- Renal osteodystrophy
**Crohn’s disease/UC**

**Ex:**

Young, thin patient with multiple abdominal scars. Could have a cutaneous fistula, ileostomy or PEG feeding. He may have Cushingoid appearances, evidence of immunosuppressive therapy, or evidence of nutritional deficiency. Look for clubbing, mouth ulcers, pyoderma gangrenosum or erythema nodosum.

A UC patient may have a lower laparotomy scar with either an ileostomy, or a repaired ileostomy site (with presumed IPAA). If jaundiced or signs of CLD – PSC!!! May have Ank Spond with quiescent Dx.

**Treatment:**

**Crohn’s:**
- Fluids
- IV steroids
- Rectal steroids if required
- LMWH – hypercoagulable
- Nutritional support
- Consideration of rescue biological therapies - Infliximab

**Ulcerative colitis**
- Topical therapy for distal disease – 5-ASA and steroids
- Systemic therapy if more severe or widespread involvement – 5-ASA and steroids
- AZA and 6-MP for maintenance off steroids
- Ciclosporin or Infliximab salvage therapy
- Pancolectomy with long rectal stump for ileal pouch anal anastomosis

**Complications of UC:**
- Colorectal cancer – 10 yrs post diagnosis then more frequently depending on risk
- Primary sclerosing cholangitis
- Pouchitis
- Anastomotic leak or stricture
- Toxic megacolon

**Extracolonic manifestations related to disease activity:**
- Uveitis
- Erythema nodosum
- Aphthous ulcers
- Acute arthropathy
- Pyoderma gangrenosum

**Extracolonic manifestations not related to disease activity:**
- Primary sclerosing cholangitis
- Ankylosing spondylitis
- Sacroiliitis
Jaundice:

Hx

Infections:
- Sore throat/Rash – EBV, CMV
- Contacts/foreign travel – Hep A or E
- IVDU – Hep C, HIV
- Transfusions – Hep B

Toxins:
- Alcohol
- Medications

Autoimmune:
- Pruritis – PBC
- Associated autoimmune phenomena

Obstructive:
- Dark urine, light stools
- Wt loss, epigastric pain – Pancreatic Ca
- Abdo pain – cholecystitis, gallstones, cholangitis
- Fever, rigors - cholangitis

Benign:
- Recurrent, familial, mild, self-resolving – Gilbert’s or Dubin-Johnson

Haemolysis:
- Hx or FHx of Sickle or thalassaemia
- Recent infection
- LUQ pain (splenic sequestration)
- Symptoms of anaemia

Examination:
- Spleen and anaemia? Haemolysis
- Hepatomegaly? Infiltration or hepatitis of some cause
- Hepatosplenomegaly with ascites – CLD with decompensation
- Pruritis and CLD – PBC
Liver transplant

Examination:
Rooftop incision with substernal extension
Gum hypertrophy if on ciclosporin
Clue as to cause

Signs of infection?
Signs of decompensation?

What are the indications for liver transplant?
Any irreversible cause of fulminant hepatic failure, decompensated cirrhosis or hepatocellular carcinoma within defined criteria

What immunosuppression are they put on?
• Calcineurin inhibitor (tacrolimus) plus purine antagonist (MMF)
• May need steroids also

What are the causes of gum hypertrophy?
• Drugs – ciclosporin, nifedipine, phenytoin
• Leukaemic invasion
• Severe gingivitis
• Granulomatous disease – sarcoid

King’s criteria
Paracetamol induced:
• pH<7.3
• INR>6.5
• Cr > 300
• Encephalopathy III or IV

Others:
• INR> 6.5
• Bili > 300
• Age
• Duration of jaundice or coma

Outcome?
• 15 year survival of 58%