Liver - Vascular Disorders - Full Clinical Guideline

Reference no.: CG-T/2013 /223

Collectively includes a number of conditions, including Budd-Chiari syndrome (BCS) and portal vein thrombosis (PVT) that can cause non-cirrhotic portal hypertension with ensuing complications. Aetiological factors can be local or systemic.

Risk factor	BCS Frequency (%)	PVT Frequency (%)
Thrombophilia		
Inherited	21	35
Acquired	44	19
Myeloproliferative neoplasm	49	21
JAK2 pos	29	16
Hormonal factors	38	44
Oral contraceptives	33	44
Pregnancy	6	0
PNH	19	0
Other systemic factors	23	n.d.
Local factors	0	21

BCS, Budd-Chiari syndrome; PVT, portal vein thrombosis; PNH, paroxysmal nocturnal haemoglobinuria; n.d, no date.

En-Vie study of patients with BCS (n=163) and PVT (n=105). Prothrombotic factors in 84% of BCS and 42% of PVT

Note JAK-2 mutation present in nearly all patients with polycythaemia rubra vera and 50% of those with essential thrombocythaemia and primary myelofibrosis.

Budd–Chiari Syndrome (BCS) - hepatic venous outflow obstruction

The level of obstruction can be located from the small hepatic venules up to the entrance of the IVC into the right atrium.

Presentation: ranges from asymptomatic to fulminant acute liver failure

- Classic symptoms - fever, abdominal pain (61%), hepatomegaly (67%) and ascites (83%) +/- lower extremity oedema, GI bleeding (5%) and encephalopathy
- 15% have co-existent PVT
- 60-80% of patients may have hepatic nodules on imaging – occur due to perfusion abnormalities, but can be difficult to distinguish from HCC.

Management:

Treatment of the underlying cause

Anticoagulation (lifelong) +/- stent/angioplasty +/- TIPSS +/- transplantation

Portal vein thrombosis (PVT)

A local cause (e.g cirrhosis, malignancy, inflammation) more commonly seen than in BCS. The prevalence of PVT is 0.6-5% in compensated liver disease and40% in patients awaiting transplantation (6 monthly USS recommended in those being assessed/ waiting for transplantation). Mortality from variceal haemorrhage is increased in those with PVT complicating cirrhosis (36 vs 16% at 6 weeks)

Acute PVT

***Presentation*:** ranges from asymptomatic to intestinal infarction

- Classic symptoms - abdominal pain (90%), systemic inflammation - fever, raised CRP/ ESR (85%)
- LFTs - generally normal or mild transaminitis
- Ascites present in 50%, but usually only evident on imaging
- **Intestinal infarction** - pain, bloody diarrhoea and metabolic acidosis (occurs if extension of thrombus into mesenteric veins leads to rapid and complete obstruction before formation of collateral circulation) – treatment laparatomy, mortality 60%
- Pylephlebitis (acute septic PVT) is characterised by fever, rigors and right upper quadrant pain. Complications include multiple liver abscesses

Chronic PVT - portal cavernoma (network of hepatopetal collaterals) fully develop in a couple of months

Presentation

- 90% present with a variceal bleed; less commonly with ascites or encephalopathy
- Hepatic decompensation in presence of cirrhosis

Management:

Treatment of the underlying cause

Surveillance and management of varices as in cirrhosis- see varices surveillance guideline

Anticoagulation – patients with acute PVT require a minimum 6 months. Recanalisation of portal vein (39%), splenic (80%) and SMV (73%) after 6-12/12. In chronic PVT consider anticoagulation on an individual case basis. In both acute and chronic PVT lifelong anticoagulation is indicated if permanent strong prothrombotic or myeloproliferative condition or intestinal ischaemia

No indication for thrombolysis (recanalisation similar to anticoagulation, increased bleeding)

Sinusoidal Obstruction Syndrome (SOS)

Sinusoidal obstruction which may extend to the level of the central vein

Causes:

- Myeloablative regimens (high dose chemotherapy +/- total body irradiation) used prior to haematopoietic stem cell transplantation (HSCT) – incidence has ↓ due to prophylaxis (defibrotide), lower radiation doses and less reliance on cyclophosphamide
- Other chemotherapeutic agents e.g cyclophosphamide
- Immunosuppressive therapy (e.g. azathioprine, 6-mercaptopurine)
- Ingestion of herbal teas made with pyrrolizidine alkaloids e.g Jamaican bush tea

Presentation:

Onset typically 10-20/7 after cyclophosphamide or >30/7 after myeloablative therapy.

- Weight gain with or without detectable ascites
- Hepatomegaly (tender)
- Jaundice and ultimately liver failure and death.

Diagnosis:

- Transjugular Liver biopsy (with HVPG measurement - supportive if > 10mmHg in patient post HSCT). Primary histological feature is centrilobular necrosis

Management:

- Supportive management is the mainstay of therapy
- Transplantation if there is a favourable prognosis relating to the original disease

(Idiopathic) non-cirrhotic portal hypertension

Requires exclusion of infiltrative disease, haematological malignancy, thrombophilia, schistosomiasis, HIV, congenital hepatic fibrosis, sarcoidosis and drug causes (e.g Azathioprine).

Presentation:

- GI haemorrhage secondary to portal hypertension
- Splenomegaly
- LFTs usually normal at initial diagnosis
- Ascites (poor prognostic sign)

Diagnosis:

- Liver biopsy – phlebosclerosis, nodular regenerative hyperplasia, sinusoidal dilatation, paraseptal shunt vessels, perisinusoidal fibrosis

Management:

- Surveillance and management of varices as in cirrhosis- see varices surveillance guideline
- Anticoagulation not recommended unless underlying prothrombotic disorder
- USS looking for PVT 6 monthly

Further reading:

EASL Clinical Practice Guidelines: Vascular diseases of the liver. Journal of Hepatology 2015

Documentation Controls

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