Acute Budd-Chiari syndrome (BCS)

Definition

Acute hepatic venous outflow obstruction not due to passive causes such as cardiac disease

- Primary BCS is defined as venous obstruction due to occlusion of any of the three major hepatic veins or the inferior vena cava, usually due to thrombosis
- Secondary BCS is due to extrinsic mechanical or invasive causes such as compression or invasion from an extrinsic lesion

BCS can be acute with acute hepatic failure, acute with rapidly worsening liver disease but not yet hepatic failure, subacute (developing over 2-3 months), and chronic manifesting with cirrhosis

Etiology

- Primary BCS: thrombosis in setting of hypercoagulable state
  - Oral contraceptive use or recent (within 2 months) pregnancy
  - Myeloproliferative disorders (e.g., polycythemia vera and essential thrombocytosis) and account for up to 25% to 50% of cases in some series
  - Systemic diseases such as systemic lupus erythematosus, antiphospholipid syndrome, inflammatory bowel disease and Behcet
  - Inherited hypercoagulable disorders such as Factor V Leiden disease, thalassemia, and protein C or S deficiency
- Secondary BCS: extrinsic mechanical or invasive causes
  - Malignancies
  - Benign space-occupying liver lesions
  - Infections such as liver abscesses

Diagnosis

- Abdominal ultrasonography with Doppler is the gold standard for the initial evaluation
  - CT scanning with contrast and MRI can also be helpful in making this diagnosis

Management

Should be undertaken using a stepwise approach including anticoagulation, angioplasty, stent, thrombectomy, thrombolysis, TIPS and liver transplantation.

- Long-term anticoagulation should be given to all patients with primary BCS and is the first-line therapy with low-molecular-weight heparin or unfractionated heparin followed by an oral anticoagulant
  - Because of the increased risk of heparin-induced thrombocytopenia, the use of unfractionated heparin is generally not recommended and may only be reserved for special situations (e.g., GFR <30 ml/min, pending invasive procedures).
- In acute cases thrombolysis can be considered
- Stenoses that are amenable to percutaneous angioplasty/stenting (short length stenoses) should be actively looked for and treated accordingly.
- TIPS insertion should be attempted in BCS when angioplasty, stenting, thrombectomy, thrombolysis are not feasible, and when the patient does not improve on medical therapy including anticoagulants.

Hyperammonemia

- Serum ammonia level is not required for the diagnosis of hepatic encephalopathy
- Ammonia level can be elevated in several nonhepatic conditions
    - Acute or chronic renal failure
    - GI bleed
    - Shock
    - Urea cycle disorders
    - Infections with urease-producing bacteria such as *Mycoplasma or Ureaplasma*
    - *Multiple myeloma*
    - *Intense muscle activity including seizures*
    - *Drugs:* valproic acid, narcotics, barbiturates, gabapentin, tacrolimus, cyclosporine, methamphetamine, carbamazepine, and others
    - Gastric or intestinal bypass surgery
- Lactulose and Rifaximin not useful when the source of increased ammonia production is not the gut

Acute hyperammonemia

- Acute liver failure
- Systemic opportunistic infections with urease-producing bacteria such as *Mycoplasma or Ureaplasma*
    - Metabolize urea as an energy source and produce ammonia
    - Posttransplant period particularly in the lung transplant (usually within the first 2 weeks to 30 days after LT surgery)
    - Tx: doxycycline and a quinolone or macrolide
- Drugs – valproic acid
- Unmasking of partial urea cycle disorders
    - Stress of surgery or infections

Treatment

- CRRT
- Intermittent HD excellent way to clear ammonia, however, rebound HA often occurs between sessions
- Neuroprotective strategies - promote urea production
    - L-arginine/L-citrulline, L-ornithine/L-aspartate, sodium benzoate, levocarnitine
- Lactulose and Rifaximin
    - Not useful in acute hyperammonemia