

ACUTE HEPATIC FAILURE IN CHILDREN

Definitions

- *Fulminant hepatic failure* (HF) - hepatic dysfunction (hepatic encephalopathy and coagulopathy) within 8 weeks of evidence of symptoms of liver disease and absence of pre-existing liver disease in any form.
- *Hyperacute/ Fulminant HF* - encephalopathy within 2 weeks of onset of jaundice
- *Subfulminant HF* - encephalopathy within 2-12 weeks of onset of jaundice
- *Subacute/ Late-onset HF* - encephalopathy later than 8 weeks to 6 months of onset of symptoms.

Salient features

- jaundice with impalpable liver or a liver of reducing size
- encephalopathy - may worsen rapidly (needs frequent review)
- bruising, petechiae or bleeding from deranged clotting unresponsive to vitamin K.
- failure to maintain normoglycaemia (which aggravates encephalopathy) or presence of hyperammonaemia
- increased intracranial pressure (fixed dilated pupils, bradycardia, hypertension and papilloedema)

Table 1. Grading of hepatic encephalopathy

| Grading of Coma level |
|---|
| Grade 1 irritable, lethargic |
| Grade 2 mood swings, aggressive, photophobia, not recognising parents, flap |
| Grade 3 sleepy but rousable, incoherent, sluggish pupils, hypertonia ± clonus, extensor spasm |
| Grade 4 comatose; decerebrate, decorticate or no response to pain |

Principles of management

Supportive Treatment

- nurse in quiet darkened room with head-end elevated at 20° with
- no neck flexion (to decrease ICP and minimise cerebral irritability).
- DO NOT SEDATE unless already ventilated
- this may precipitate respiratory failure and death.
- maintain blood glucose between 6-9 mmol/l using *minimal fluid volume* (40-60 ml/kg/day crystalloid) with high dextrose concentrations e.g. 10-20%. Add Potassium as necessary.
- check dextrostix 2 - 4 hourly.
- strict monitoring of urine output and fluid balance. Catheterise if necessary.
- check urinary electrolytes, serum urea, creatinine, electrolytes and osmolarity.
- frequent neurological observations (1-4 hourly).
- maintain oxygenation with facial oxygen.
- give vitamin K to attempt to correct prolonged PT. If frank bleeding (GIT/oral) occurs, consider prudent use of FFP or IV cryoprecipitate at 10 ml/kg.
- prophylactic ranitidine plus oral antacid to prevent gastric, duodenal ulceration.
- full septic screen (excluding LP) on admission, CXR. Treat sepsis aggressively, monitoring levels of aminoglycosides frequently.
- stop oral protein initially. Gradually reintroduce 0.5-1g/kg/day.
- lactulose to produce 3-4 loose stools per day.
- ***strict fluid balance is essential** - aim for urine output < than 0.5 ml/kg/hour.
- consider N-acetylcysteine.

Clinical Pearls In a comatose patient:

- in the presence of *sudden coma*, consider intracranial bleed - request a CT Brain.
- patients in Grade 3 or 4 coma require mechanical ventilation to maintain normal cerebral perfusion pressure.

Table 2. Causes of hepatic failure

| |
|---|
| Infection |
| hepatitis A, B, non A-non B, CMV, leptospirosis, Dengue |
| Drugs |
| carbamazepine, valproate, paracetamol, halothane |
| Ingested toxins |
| mushrooms, <i>Amanita phalloides</i> |
| Metabolic |
| fructosaemia, galactosaemia, tyrosaemia, Wilson's disease |
| Ischaemic shock |
| gram negative septicaemia, Budd Chiari syndrome |
| Tumour |
| histiocytosis, lymphoproliferative disorder |

Table 3. Fluid management in liver failure

| | Normal Liver Function | Liver failure |
|--|-----------------------|--|
| Volume given if no dehydration and losses are not abnormal | | |
| Body Weight | | |
| < 10 kg | 120-150 ml/kg/day | 60-80 ml/kg/day |
| 10-20 kg | 90-120 ml/kg/day | 40-60 ml/kg/day |
| > 20 kg | 50-90 ml/kg/day | 30-50 ml/kg/day |
| Fluid type | Dextrose 4 – 5 % | Dextrose ≥ 10% (adjust according to Destrostix readings) |
| Potassium | 1 - 3.5 mmol/kg/day | NIL WHILE ANURIC |
| Sodium | 1.5 - 3.5 mmol/kg/day | NIL ADDED |
| Other Fluids | Albumin 20% 5 ml/kg | Albumin 20% 5 ml/kg |
| For transfusion | FFP 10-20 ml/kg | FFP 10-20 ml/kg |
| Blood volume (ml) = No. of grams to raise Hb by x body weight in kg x F Where F = 6 for whole blood, F = 4 for packed cells | | |