

NHS Children's Acute Transport Service



Clinical Guidelines

Acute Liver Failure

Document Control Information

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1. Definitions

• **Acute Liver Failure:**

Rare multisystem disorder involving massive hepatic necrosis associated with severe impairment of liver function with or without encephalopathy in a patient with no recognised underlying chronic liver disease. In the absence of encephalopathy the severity of liver impairment is best assessed by the degree of coagulopathy.

Paediatric Acute Liver Failure Group definition: Coagulopathy with INR ≥ 1.5 with encephalopathy or INR ≥ 2 without encephalopathy due to a liver cause, not correctable by intravenous vitamin K.

• **Fulminant Liver Failure:**

Onset of hepatic encephalopathy and coagulopathy within 8 weeks of onset of liver disease, in the absence of pre-existing liver disease in any form.

2. Assessment

- Aetiology of liver failure

Neonatal:

- neonatal haemochromatosis
- viral hepatitis: HSV
- HLH
- metabolic disorders

Children:

- infective: non A-E hepatitis, parvovirus, adenovirus
- septicaemia
- toxic: paracetamol, sodium valproate, carbamazepine, isoniazid, rifampicin, halothane
- infiltrative: leukaemia, HLH
- autoimmune
- ischaemic: Budd Chiari syndrome, shock
- metabolic
- cryptogenic

- Clinical features
 - Jaundice
 - Altered conscious level (encephalopathy)
 - Ascites, effusions
 - GI haemorrhage (variceal haemorrhage in portal hypertension)
 - Hepatomegaly (may shrink with deterioration)
 - Systemic collapse (poor prognosis)

- Biochemical findings
 - Hepatic injury:
 - raised transaminases (ALT, AST,GGT)
 - Hepatic dysfunction:
 - Coagulopathy (PT > 20sec) not correctable with vitamin K
 - Hypoglycaemia
 - Hypoalbuminaemia
 - Increasing bilirubin
 - Hyperammonia and encephalopathy
 - High lactate

Investigations requested should therefore include:

Bilirubin (total, conj), AST, ALT, GGT, Alb, NH₃, Lactate, Glucose
FBC, clotting (including PT/INR, APTT, Fib)

- Common complications
 - Cerebral oedema, raised intracranial pressure
 - Sepsis
 - Coagulopathy
 - Respiratory failure, aspiration
 - Renal failure
 - MODS
 - Adrenal insufficiency
 - Electrolyte disturbances (low Na, K, Ca, Mg, phosphate)

3. Indications for transfer to King's College Hospital (KCH)

- Altered level of consciousness
- Elevated INR >1.6 in absence of DIC and unresponsive to Vitamin K and FFP, with elevated or decreasing transaminases
- Hypoglycaemia
- Jaundice
- Metabolic acidosis
- Hyperlactataemia

Not all of these indications require intensive care transfer by CATS.

Liver failure requires specialised management and advice should not be given to referral hospitals without prior discussion with the KCH Liver and PICU consultant and the CATS team.

4. Initial management

- High flow oxygen Secure and maintain airway.
- Obtain venous access. The preferred route for central access is femoral.
- Treat coagulopathy with IV vitamin K 2-10mg. Avoid the use of blood products unless actively bleeding, requiring invasive procedures or severe coagulopathy (PT>60 sec or INR > 4.0). In these circumstances treat with FFP and cryoprecipitate and aim for a platelet count of $>50 \times 10^9/L$
- Monitor blood glucose and maintain between 4-8mmol/l. This may require 10-20% dextrose infusion. (NB: 20% dextrose should be given centrally)
- 0.9% NaCl +10% Dextrose should be used for maintenance. Fluid restrict all patients to 2/3 calculated maintenance.
- Site nasogastric tube or orogastric tube (occasionally avoided in coagulopathy)
- Institute regular neuro observations (every 15 minutes).
- Start antibiotics and antifungals in all cases and additionally parenteral high dose acyclovir (60 mg/kg/day) in all cases of neonatal acute liver failure.
- Commence N-acetylcysteine (Parvolex) in paracetamol poisoning or if instructed by the KCH team (100 mg/kg/day as a continuous infusion)

Table: Hepatic Encephalopathy Grading

Grade 0	Clinically normal mental status but minimal changes in memory, concentration, intellectual function, and coordination
Grade 1	Mild confusion/anxiety, disturbed or reversal of sleep rhythm, shortened attention span, slowing of ability to perform mental tasks (simple addition or subtraction). In young children, irritability, altered sleep pattern, unexplained bursts of excessive crying.
Grade 2	Drowsiness, confusion, mood swings with personality changes, inappropriate behaviour, intermittent disorientation of time and place, gross deficit in ability to perform mental tasks. In young children, excessive sleepiness, inability to interact with or recognise parents, lack of interest in favourite toys and activities.
Grade 3	Pronounced confusion, delirious but arousable, persistent disorientation of time and place, hyperreflexia with a positive Babinski's sign.
Grade 4	Comatose with or without decerebrate or decorticate posturing, response to pain present (IVa) or no response to pain (IVb).

5. Indications for intubation

- \geq Grade 2 encephalopathy
- Raised intracranial pressure
- Rapidly deteriorating course
- Respiratory failure
- Cardiovascular collapse
- Portal hypertension with variceal haemorrhage

Oral intubation is preferred, with a cuffed ETT, due to risk of bleeding and aspiration.

6. Management following intubation

- Aim to oxygenate ($\text{PaO}_2 > 10 \text{ kPa}$) and maintain normocarbida ($\text{PaCO}_2 4.5- 5.3$)
- If raised ICP suspected, treat as such (see guideline on traumatic head injury). Consider the use of 2.7% saline (aim for Na 145mmol/l)
- Femoral central access if vasoactive agents or high concentration dextrose infusions are required.
- Noradrenaline is the vasoactive agent of choice.
- Consider IV hydrocortisone 1-2 mg/kg 6 hourly if adrenal insufficiency suspected. Draw blood for baseline cortisol first but do not wait for assay.
- **If intubated for any grade of encephalopathy – early neuroprotection (same principles as for TBI)**

7. Transport considerations

- Paralyse and sedate.
- Monitor pupils.
- Monitor arterial gases, glucose and lactate levels closely.
- Keep normal saline fluid boluses ready during journey. Keep 2.7% saline ready to give as a bolus if required
- For non-intubated patients: no sedation (risk of loss of airway), avoid any unnecessary pain and stimulation.

Acute Liver Failure patients rarely have varices - hence variceal bleeding is rare

In chronic liver disease or those with acute on chronic liver failure, portal hypertension and variceal bleeding might pose particular challenge

For uncontrolled variceal haemorrhage

- Correct coagulopathy
- Give Vitamin K
- Ensure packed cells given and more available for transfer, as well as other blood products
- Commence octreotide infusion (mix 500mcg in 40mls 0.9% NaCl, run at 2-4 ml/hr)
- Consider placing Sengstaken tube (if appropriately trained)
- Use an oral cuffed tube for intubation

Discuss with CATS consultant and KCH consultant on call