

NHS **CATS**
Children's Acute Transport Service



Clinical Guidelines

Acute Liver Failure

Document Control Information

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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 (PALF) Study Group definition:

Coagulopathy with INR ≥ 1.5 with encephalopathy or INR ≥ 2 with or without encephalopathy due to a liver cause, not correctable by intravenous vitamin K.

Assessment

Aetiology of liver failure

Neonatal:

- Gestational alloimmune liver disease (previously known as neonatal haemochromatosis)
- Viral hepatitis: HSV
- Haemophagocytic Lymphohistiocytosis (HLH)
- Metabolic disorders

Children:

- Infective: non-A-E hepatitis, parvovirus, adenovirus
- Septicaemia
- Toxic/Drug induced: 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)
- Cardio-vascular collapse (poor prognosis)

Biochemical findings

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

Baseline investigations: bilirubin (total, conjugated), AST, ALT, GGT, Alb, NH₃, lactate, glucose, FBC, clotting (including PT/INR, APTT, fibrinogen).

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)

Indications for transfer to King's College Hospital (KCH), London

- Altered level of consciousness
- Elevated INR >1.5 in the absence of DIC and unresponsive to Vitamin K with elevated or decreasing transaminases
- Hypoglycaemia
- Jaundice
- Metabolic acidosis
- Hyperlactataemia

Not all these indications require intensive care transport 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.

Initial management

- Secure and maintain airway if required, fluctuating level of consciousness needs to be discussed with CATS/KCH teams as intubation/ventilation may be required for transport.
- Obtain venous access. The preferred route for central access is femoral.
- Treat coagulopathy with IV vitamin K. 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. Treat hypoglycaemia with 3mls/kg bolus of 10% glucose. This may also require 10-20% glucose infusion. (NB: 20% glucose should be given centrally) making sure that glucose is given with normal saline and not as hypotonic plain dextrose solution.
- 0.9% sodium chloride + 10% glucose 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 intravenous high dose aciclovir (60mg/kg/day) in all cases of neonatal acute liver failure.
- Commence N-acetylcysteine (Parvolex) in paracetamol poisoning or if instructed by the KCH team (100mg/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 rousable, 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).

Indications for intubation

- > Grade 2 encephalopathy
- Raised intracranial pressure
- Rapidly deteriorating course
- Respiratory failure
- Cardiovascular instability
- Portal hypertension with variceal haemorrhage (though rare in ALF)

Avoid nasal intubation due to risk of bleeding and aspiration. Cuffed ETT should be used.

Management following intubation

- *If intubated for any grade of encephalopathy – early neuroprotection (same principles as for TBI).*
- Aim to oxygenate ($\text{PaO}_2 > 10 \text{ kPa}$ /Sats > 94%) and maintain normocarbida (PaCO_2 4.5- 5.3 kPa)
- Consider the use of 2.7% sodium chloride (aim for Na 145-150 mmol/l)
- Central access (preferably femoral) if vasoactive agents or high concentration glucose 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.

Transport considerations

- Sedate and muscle relax.
- Monitor pupils.
- Monitor arterial gases, glucose and lactate levels closely.
- Have 0.9% sodium chloride fluid boluses ready during journey. Have 3 ml/kg 2.7% sodium chloride 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.
- Ongoing neuro-protection (as mentioned above) for children intubated for hepatic encephalopathy of any grade.

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 a particular challenge.

For uncontrolled variceal haemorrhage

- Activate major haemorrhage protocol.
- Correct coagulopathy; consider tranexamic acid.
- Give Vitamin K
- Ensure packed cells given and more available for transport, as well as other blood products.
- Commence octreotide infusion (mix 500mcg in 40mls 0.9% sodium chloride, 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.

Reference:

BSPGHAN. *Acute Liver Failure Guidelines*. March 2023. Available at: <https://bspghan.org.uk/wp-content/uploads/2023/08/acute-liver-failure-guidelines.pdf>