

Clinical Guidance

Paediatric Critical Care: Paediatric Acute Liver Failure (PALF)

Summary

This guideline is for staff to use to provide guidance regarding the management of children with acute liver failure on Paediatric critical care and on retrieval. It discusses areas for considerations and offers treatment advice.

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Author(s)	Jon Lillie, PICU Consultant Jennie Lambert, PICU Consultant	
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Relevant external law, regulation, standards		
<p>This clinical guideline has been produced by the South Thames Retrieval Service (STRS) at Evelina London for nurses, doctors and ambulance staff to refer to in the emergency care of critically ill children. This guideline represents the views of STRS and was produced after careful consideration of available evidence in conjunction with clinical expertise and experience. The guidance does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient.</p>		
Date	Change details, since approval	Approved by
January 2021	Minor formatting changes, references updated, mannitol removed and 2.7% sodium chloride dose standardised throughout, fluid bolus 20mL/kg to 10mL/kg and dopamine removed as per UK resus council guidance 2021 Antibiotic guidance altered as per PIID/ ASC advice.	ELCGC February 2022 ASC September 2022

Glossary:

LFT–liver function test, **FBC**–full blood count, **U&E**–urea & electrolytes, **INR**–international normalised ratio, **WBC**–white blood cell, **AST**–aspartate aminotransferase, **GGT**–gamma-glutamyl transferase, **CXR**–chest x-ray, **FFP**–fresh frozen plasma, **ICP**–increased intracranial pressure **GCS**–Glasgow Coma Scale

Definition:

Acute loss of hepatocellular function secondary to hepatocellular injury or death. Devastating illness, with progressive, rapid, multisystem organ failure.

Defining clinical features:

- Biochemical evidence of liver injury (no chronic impairment)
- Coagulopathy (INR>2) OR Coagulopathy (INR>1.5) AND encephalopathy

Causes:

Unknown (>50%), infective, metabolic, drugs, auto-immune, vascular, ischaemic, infiltrative

Poor Prognostic Indicators:²

- INR >4 despite 1 dose of vitamin K
- Age <5 year, especially neonates
- Bilirubin \geq 235 micromol/L
- Elevated lactate
- WBC $>9 \times 10^9/L$
- Grade 3/4 coma in Paracetamol overdose (see right)

NB: Fixed dilated pupils may be reversible

Grading Encephalopathy²

- 1: Confused, mood changes
- 2: Drowsy, inappropriate behaviour
- 3: Stuporous, sleepy but rousable
- 4a: Comatose but rousable to pain
- 4b: Deep coma, unrousable (Encephalopathy rare in children)

**NB: It is very difficult to standardise assessment of encephalopathy in paediatrics
Deteriorating GCS is always concerning**

*****Transfer early – before uncontrolled bleeding & encephalopathy*****

King's Liver Registrar: 020 3299 7812 ext. 37812 (09:00-17:00) Out of hours: 07866 792368

Kina's PICU: 0203 2993660 Rays of Sunshine Ward: 0203 2993577

Resuscitation and routine management:**A→E assessment, frequent re-evaluation:**

- Oxygen as required to titrate sats 94-98%
- Indications for intubation: pulmonary oedema, fluid refractory shock (>60ml/kg), encephalopathy Grade 2 and above
- IV fluid boluses in 10 mL/kg aliquots, titrated to CVS response
- Inotropes if fluid-refractory shock
- Consider central line early, remember bleeding risk (optimise coag)
- 60% maintenance with 0.9% sodium chloride and 5%/10% glucose
- **AVOID** hypotonic fluids (↑risk hyponatraemia & cerebral oedema)
- Site urinary catheter to monitor urine output (risk of renal failure)
- Insert nasogastric tube or orogastric tube if significant coagulopathy
- Cover for sepsis/ viral: IV cefotaxime in all patients. In neonates, add IV aciclovir
- Sedate with morphine/ midazolam infusion as required if intubated
- Discuss with pharmacy regarding drugs metabolised by liver

Baseline investigations:

FBC, glucose, U&E, coagulation, blood gas, lactate, LFTs (including conjugated bilirubin, AST, GGT), ammonia, blood cultures & CXR

*****Regular re-assessment of coagulation profile & glucose**

Intubation – indications & strategy:

- Consider intubation if encephalopathy Grade 2 or more
- 3 mL/kg 2.7% sodium chloride pre-induction if encephalopathic
- Ketamine, fentanyl and rocuronium as per STRS practice
- Use oral tracheal tube (coagulopathy)

Ventilation strategy:

- PEEP \geq 5cm H₂O for SpO₂ 94-98%
- ETCO₂ 4-5cm H₂O
- Hyperventilate if ICP crisis

Fluid refractory shock:

- Noradrenaline is first line vasopressor (warm shock)³
- Add adrenaline if poor cardiac function
- Consider 2mg/kg IV hydrocortisone

Paracetamol overdose (see local guideline)

Advice: **GSTT toxicology** or **TOXBASE** 0344 8920111

N- acetyl cysteine (NAC) therapy:

- 1) **Single point ingestion** – plot timed paracetamol level on treatment nomogram, based on risk stratification
- 2) **Staggered ingestion** – treat if $>150\text{mg/kg/24h}$ ingested (threshold lowers to 75mg/kg/24h if high risk e.g. chronic diseases, malnutrition, therapeutic use of inducers, such as phenytoin, chronic alcohol excess).

***NAC should be continued** at 150mg/kg/24hrs in patients with PALF, renal failure or deranged coagulation.

Hypoglycaemia in PALF:

Loss of the liver's ability to control glucose homeostasis.

Therapy must maintain normoglycaemia (4-7mmol/L)

If blood glucose <4mmol/L:

- Initial bolus 5mL/kg 10% glucose
- Increase glucose concentration in maintenance fluids (up to max of 25%) to maintain stable blood glucose – avoid repeated boluses as can cause rebound hypoglycaemia
- NB: need central access if using $>12.5\%$ glucose

Bleeding in PALF:

Loss of Liver's central role in haemostasis, including inability to produce clotting factors & platelet dysfunction

Treatment:

- IV vitamin K (phytomenadione) 1mg/kg, maximum 10 mg
- FFP 10 mL/kg, Platelets 15 mL/kg, Cryoprecipitate 5mL/kg
- Consider recombinant factor VIIa 80 micrograms/kg in persistent haemorrhage² (Fibrinogen must be $>1\text{g/L}$)

Management of GI Haemorrhage:

- IV pantoprazole
- Consider variceal bleed - get surgical consultation - place Sengstaken tube if available
- **Octreotide** Bolus 1 microgram/kg then infusion 1 to 3 micrograms/kg/h (start at DGH if suspected variceal bleed)

Encephalopathy in PALF:

Multi-factorial - associated with failure of the liver to remove toxins from the blood, deranged cellular osmolarity & cerebral oedema

Management:

- Intubate if encephalopathy Grade 2 or more
- Maintain normocapnia (ETCO₂ 4-5cm H₂O), normothermia and normoglycaemia (see above)
- Treat seizures: load IV phenytoin or levetiracetam (APLS)
- Target mean arterial pressures: 60mmHg if <4 years and 65 mmHg if >4 years
- Give osmotherapy 2.7% sodium chloride (1-2 mL/kg) If raised ICP (assume if hypertensive, bradycardic or dilated pupils) – bolus 3mL/kg 2.7% sodium chloride and review
- If ICP crisis – hyperventilate

Renal Impairment in PALF:

Many potential causes e.g. shock, hepatorenal syndrome

****Early consultation with renal team advised****

- Maintain urine output $>1\text{mL/kg/h}$: standard fluid challenge, consider furosemide depending on cause
- Correct low potassium/ calcium/ magnesium
- NB uraemia in renal impairment can impair platelet function & worsen coagulopathy