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Paediatric Acute Liver Failure CoMET Guideline

This guideline is for use by healthcare staff, at CoMET undertaking critical care retrieval, transport and stabilization of children, and young adults.

CoMET is a Paediatric Critical Care Transport service and is hosted by the University Hospitals of Leicester NHS trust working in partnership with the Nottingham University Hospitals NHS Trust.

The guidance supports decision making by individual healthcare professionals and to make decisions in the best interest of the individual patient.

This guideline represents the view of CoMET, and is produced to be used mainly by healthcare staff working for CoMET, although, professionals, working in similar field will find it useful for easy reference at the bedside.

We are grateful to the many existing paediatric critical care transport services, whose advice and current guidelines have been referred to for preparing this document. Thank You.

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Education and Training

1. Annual Transport team update training days
2. Workshops delivered in Regional Transport Study days/ Outreach

Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Incident reporting	Review related Datix	Abi Hill – Lead Transport Nurse abi.hill@uhl-tr.nhs.uk	Monthly	CoMET Lead Governance Meeting
Documentation Compliance	Documentation Audit	Abi Hill – Lead Transport Nurse abi.hill@uhl-tr.nhs.uk	3 Monthly	CoMET Lead Governance Meeting

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Paediatric Acute Liver Failure (PALF)

Definition

Biochemical evidence of acute liver injury with no chronic liver impairment

Coagulopathy not corrected by IV Vitamin K where:-

- PT ≥ 15 seconds or INR ≥ 1.5 with encephalopathy
- PT ≥ 20 seconds or INR ≥ 2 with/or without encephalopathy

Causes: - unknown (>50%), infective, metabolic, drugs, autoimmune, vascular, ischaemic, infiltrative

NB *It is difficult to standardize assessment of paediatric encephalopathy. However deteriorating GCS is always concerning*

Early Discussion with Local Paediatric Hepatology team is required if you are suspecting PALF

Birmingham Children's Hospital Contact via 0121 333 9999 (switchboard and then ask for hepatology team)

Leeds 01132432799 (switchboard and then ask for Paediatric Consultant Hepatologist)

King's College Hospital in-hours 02032999000 ext 37812 out of hours 07866792368

Routine Management

• Initial management in Figure 1, requires frequent re-evaluations

• Fluids at 2/3rd maintenance of 0.9% sodium chloride and 10% glucose

• Avoid hypotonic fluids (as increased risk of hyponatraemia and cerebral oedema)

• Site Urinary catheter by a competent and experienced practitioner due to bleeding risk to monitor urine output aiming >0.5mls/kg/hour (risk of renal failure)

• Insert nasogastric tube or orogastric tube if significant coagulopathy

• Cover for sepsis/viral causes:-

O IV Cefotaxime in all patients (empirical antimicrobials should be used in the context of patients microbiological and allergy history)

O IV aciclovir in neonates and consider in other patients

• Start IV Omeprazole 2mg/kg

• If intubated sedate with morphine/midazolam infusion as required

• Discuss with pharmacy regarding drugs metabolized by liver

Baseline investigations

FBC, glucose, U&Es, coagulation including fibrinogen, blood gas, LFTs, conjugated bilirubin, AST, GGT, ammonia, paracetamol and toxicology, blood cultures, CXR. Consider CT head if encephalopathy

****Regular re-assessment of coagulation profile and**

Hepatic Encephalopathy

	Clinical	Neurological signs
Grade 1	Confused / euphoric irritability, excessive crying, sleep reversal inattention to task	Delayed response (<4y), tremor, apraxia, impaired handwriting, slight asterixis, reflexes normal or hyperreflexia
Grade 2	Drowsy / restless, mood swings, inappropriate behaviour, excessive sleepiness, no interaction with parents, lack of interest in toys	Progressive decrease in response (<4y), ataxia, dysarthria, asterixis present, reflexes normal or hyperreflexia
Grade 3	Pronounced confusion, delirious but arousable, somnolence, stupor, combativeness	Progressive decrease in response (<4y), positive Babinski, rigidity, asterixis present, hyperreflexia
Grade 4	Comatose, arouses with painful stimuli or no response	Decerebrate or Decorticate posturing, absent reflexes

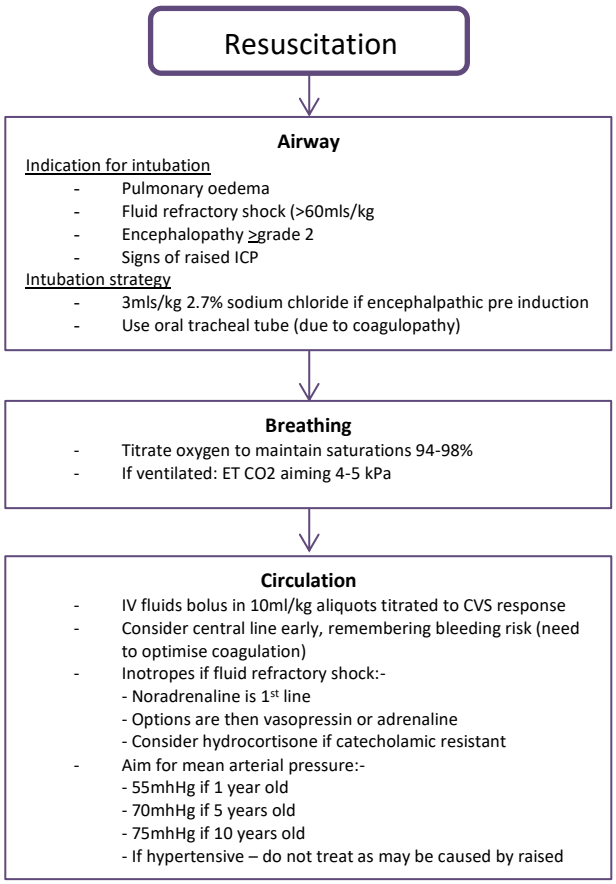

Fig 1: showing initial resuscitation in terms of A to C

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Paediatric Acute Liver Failure (PALF) Troubleshooting

Hypoglycaemia in PALF

Loss of the liver's ability to control glucose homeostasis

Must maintain normoglycaemia (4-7mmol/l)

If blood sugar <4mmol/l:-

- Initial bolus of 5mls/kg of 10% glucose
- Increase glucose concentration in maintenance fluids (up to max of 25%) - Central access required if using ≥12.5% dextrose
- Avoid repeated boluses as this can cause rebound hypoglycaemia

Renal Impairment in PALF

Many potential causes i.e shock, hepatorenal syndrome

Early consultation with renal team advised

- Consider fluid bolus 10mls/kg and/or furosemide 1-2mg/kg IV (max40mg)
- Maintain urine output > 1ml/kg/hr
- Correct low potassium/calcium/magnesium

Be aware that uraemia can cause platelet dysfunction and worsen coagulopathy

Encephalopathy in PALF

This is multi-factorial and is associated with failure of the liver to remove toxin from the blood, deranged cellular osmolarity and cerebral oedema

Management

- Intubate if Grade 2 Encephalopathy or more
- If intubated, then requires neuroprotection measures (i.e head up 30 degree and in midline, normoglycaemia, avoid seizures, aim PCO_2 4-5kpa)
- Treat seizures
  - If not intubated – avoid benzodiazepines due to risk of loss of airway and respiratory depression. Load with IV levetiracetam
  - If Intubated as per APLS
- Target mean arterial pressures at 50th centile, >5th centile

Age	>5th centile	50th centile
1 month	40	55
1 year	50	70
>5 years	55	75

- If raised ICP (hypertensive, bradycardic or dilated pupils) – bolus 3mls/kg 2.7% Sodium chloride and review
- If ICP crisis – hyperventilate

Perform CT head if signs of ICP and encephalopathy requiring intubation to rule out bleeding

Bleeding in PALF

Loss of liver's central role in haemostasis, including inability to produce clotting factors and platelet dysfunction.

This is an important prognostic marker and should not be treated without discussion with hepatologist

Indications for treatment:

- **Active severe bleeding**
- **Requirement for invasive procedure**
- **If already listed for transplant**

Treatment:-

- IV Vitamin K 1mg/kg max 10mg
- FFP 10mls/kg and platelets 15mls/kg cryoprecipitate 5ml/kg
- Aiming Platelets >50, INR <2 fibrinogen > 1g/l
- Consider recombinant factor VIIa 80micrograms/kg in persistent haemorrhage (Fibrinogen must be > 1g/l)

Management of GI Haemorrhage

- Consider variceal bleed – get surgical consultation
- Treatment as above, including RBCs
- Will require blood products for transport
- Octreotide bolus 1microgram/kg then continuous infusion of 1-3micrograms/kg/hr

Consideration of Sengstaken tube (see separate guideline)

Paracetamol Overdose

- Refer to local guidelines for initial management
- If further advice required consult Toxbase or NPIS on 0344892 0111
- If NAC started for paracetamol overdose then should be continued in patients with PALF

Transport Considerations:

- To have free lumen/cannula present in order to give bolus as required
- Have crystalloid fluid boluses and 2.7% sodium chloride ready during the transfer to give if required
- If concerns regarding GI haemorrhage, having blood products available for transfer
- For non-intubated patients
  - No sedation (as risk of loss of airway)
  - Avoid any unnecessary pain and stimulation
- For intubated patients:
  - Sedate and muscle relax
  - Monitor pupils and signs of ICP

On-going neuro-protection (same principles as in TBI)

References

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