



PAEDIATRIC ACUTE CARE GUIDELINE

Hyperammonaemia

Scope (Staff):	All Emergency Department Clinicians
Scope (Area):	Emergency Department

This document should be read in conjunction with this DISCLAIMER
<http://kidshealthwa.com/about/disclaimer/>

Hyperammonaemia

Hyperammonaemia can be defined as:

- Premature neonate - $\text{NH}_4 > 150\mu\text{mol/L}$
- Term neonate - $\text{NH}_4 > 100\mu\text{mol/L}$

Background

- Elevated plasma ammonia is a **medical emergency**
- Consider hyperammonaemia in the differential diagnosis of **any sick neonate**
- The neurological outcome of affected neonates is directly related to the duration of hyperammonaemic coma
- Contact the Emergency Department Consultant urgently if the patient is unwell

History

- Clinical signs of hyperammonaemia are non specific but can include tachypnoea, seizures and encephalopathy

Investigations

Ammonia (x2)	Free flowing, consider arterial sample if difficult. Place on ice and transport urgently to lab.
Venous blood gas	Respiratory alkalosis in Urea Cycle Defects, metabolic acidosis in Organic Acideamias
Lactate	Raised in Organic Acideamias, Fatty Acid Oxidation defects and Urea Cycle defects with circulatory collapse

Liver Function	Deranged in liver failure, Organic Acidaemias and Fatty Acid Oxidation defects
Clotting	Deranged in liver failure, Organic Acidaemias and Fatty Acid Oxidation defects
Urine Ketones (dipstick)	Low in Fatty Acid Oxidation defects and Urea Cycle defects. Raised in Organic Acidaemias.
Plasma Amino Acids	
Urine Amino Acids	
Urine Organic Acids	
Plasma Acylcarnitines	Can also be done on the Guthrie card

Differential diagnoses

- Spurious
 - Sample haemolysed, not collected on ice, or delayed separation
- Hepatic liver failure/impairment
- Metabolic
 - Urea Cycle defects (UCD)
 - Organic Acidaemias (OA)
 - Fatty Acid Oxidation defects (Fatox)
- Transient hyperammonaemia of the newborn (due to open ductus venosus)

Management

- Elevated plasma ammonia is a **medical emergency**
- Do not delay treatment whilst awaiting results of further investigations

Initial management

Stop all enteral feeds
Promote anabolism : • Start intravenous 10% Dextrose to ensure glucose infusion of 8-10mg/kg/minute, aiming for a blood glucose level of 4-8 mmol/L
Add Insulin infusion of 0.05U/kg/hr if blood glucose > 15mmol/L. Do not turn down the rate of 10% Dextrose (remember the aim is to stop catabolism and this can only be done by giving lots of calories).
If ammonia > 150 µmol/L, commence: • Sodium Benzoate 250mg/kg/day as a continuous IV infusion and • Carnitine 100mg/kg/day as a continuous IV infusion • These products are now available as concentrated solutions in ampoules in the hyperammonia kit on 6B (Neonatal Unit)
If ammonia > 300µmol/L • Need to prepare for probable haemodiafiltration • Contact PICU Consultant to discuss admission and further management
Remember: It can take some time to get appropriate central venous access and commence haemodiafiltration so ALL other measures to lower plasma ammonia must be initiated as soon as possible.
In addition, consider arginine 350/kg/day as a continuous infusion

If boluses of fluid are required:


- Use 0.9% saline
- Remember: 4.5% Human Albumin solution and Fresh Frozen Plasma contain protein

- In the event of imminent death, an antemortem liver biopsy and a skin biopsy should be obtained, to assist with diagnosis
- Contact the biochemistry laboratory to arrange
- 2mls EDTA whole blood should also be collected for potential genetic studies
- Finally, consider carefully whether to perform a post-mortem, even if liver and skin biopsies have been taken

Tags

ammonia, arginine, carnitine, encephalopathy, fatty acid oxidation defect, hyperammonaemia, nh4, organic acidaemias, seizures, tachypnoea, urea cycle defects

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