# Guideline Outline

Clinical suspicion of a metabolic disorder/hyperammonemia

- A to E
- Blood sugar, gas, FBC, U&E, LFT, CRP, and cultures
- Send URGENT AMMONIA: venous sample, on ice, alert lab prior to taking sample to prevent processing delays
- Treat for possible sepsis with broad spectrum IV antibiotics and IV acyclovir if concerns about herpes infection
- STOP FEEDS and start IV fluids containing 10% dextrose

Ammonia ≥ 150 µmol/L

Repeat ammonia. Call lab to ensure no delays in processing sample.

Ammonia < 200

Ammonia ≥ 200

Discuss with on call metabolic consultant & inform Embrace if clinically deteriorating. Consider alternate causes of hyperammonaemia (HSV, drugs, sepsis, etc) or shock (congenital heart disease in a neonate)

Ammonia ≥ 200 µmol/L

- Contact Metabolic Consultant
- Start metabolic infusions **WITHIN 30 minutes** of decision to treat (pg 3)
- 2nd peripheral venous access

Repeat ammonia and gas one hour after starting metabolic infusions

**Ammonia rising despite treatment**

**Ammonia ≥ 400 µmol/L**

## HYPERAMMONAEMIA IS A TIME CRITICAL MEDICAL EMERGENCY

**AMMONIA >400 µmol/L RESISTANT TO PHARMACOLOGICAL TREATMENT MUST START HEAMOFILTRATION WITHIN 6 HOURS OF IDENTIFICATION**

- Start metabolic infusions **WITHIN 30 minutes** of decision to treat (see pg 3)
- Contact anaesthetic SpR/Cons for assessment
- Intubate and ventilate after discussions with PICU consultant via Embrace
- Consider carglumic acid after discussions with metabolic consultant
- Send repeat ammonia pre-transfer

***DO NOT DELAY TIME CRITICAL TRANSFER TO PICU!***

Embrace: 0114 268 8180Metabolic consultant on call (at Royal Manchester Children's Hospital): 01612761234

***TIME IS BRAIN*** WHEN MANAGING HYPERAMMONAEMIA. AMMONIA IS NEUROTOXIC AND THE RISK OF PERMANENT NEUROLOGICAL DAMAGE AND DEATH IS DIRECTLY RELATED TO THE **DEGREE** AND **DURATION** OF AMMONIA PEAK. TREAT THIS WITH THE UTMOST URGENCY.

Reference: NWTS Guideline for the Management of Neonatal and Paediatric Hyperammonaemia. 2018.