Management of Neonatal & Paediatric Hyperammonaemia

Title:	**Management of Neonatal and Paediatric Hyperammonaemia**
Version:	**3**
Supersedes:	Version 2
Application:	The guideline is intended for use by any hospital team caring for infants, children and young people under 16 years age across the Paediatric Critical Care Network in the North-West (England) & North Wales region.

Originated /Modified By: Designation:	**Version 1 Authors:** Rachael Barber, PICM Consultant, NWTS and RMCH; Adam Nicholls, Paediatric SpR, RMCH; Suzy Emsden, PICM Grid Trainee AHCH; Sharryn Gardner, EM Consultant, Southport & Ormskirk Hospitals NHS Trust; Adam Sutherland, Metabolic Pharmacist, RMCH; Beth Jameson and Simon Jones, Consultants in Metabolic Medicine, RMCH **Version 2: Authors:** Rachael Barber, PICM Consultant, NWTS and RMCH; Andrew Taylor, Paediatric Pharmacist, Alder Hey; Ian Dady, Consultant Neonatologist and Clinical Lead, GMNETS; Sarah McBride, Neonatal Pharmacist, St Mary's **Version 3: Authors:** Benedict Rafferty PICM Grid trainee Alder Hey/Belfast, Kate Parkins, PICM Consultant, NWTS Andrew Morris & Simon Jones Consultants in Metabolic Medicine, RMCH; Sophina Mahmood, Paediatric Pharmacist, PCC ODN / RMCH Rebecca Hinton, Paediatric Pharmacist, PCC ODN / RMCH
Amendments	Algorithm changed, clarifying thresholds for treatment Changes to ammonia sampling information Clarification of glucose containing fluids used for metabolic patients Clarification of urgency of transfer and who should transfer
Reviewed by:	North-West (England) and North Wales Paediatric Transport Service (NWTS) North-West Neonatal Network North-West & North Wales Paediatric Critical Care ODN
Ratified by:	1. North-West (England) & North Wales Paediatric Critical Care Operational Delivery Network 2. RMCH (Host Trust): Paediatric Policies & Guidelines & Pharmacy & Medicines Management Committees
Date of Final Ratification:	27th January 2025
Issue / Circulation Date:	Version 3 / circulated to PCC ODN 11.04.24 & Neonatal ODN 15.04.24 (for comments
Circulated by:	PCC, SiC & LTV ODN + Neonatal ODN
Dissemination and Implementation:	Via PCC & Neonatal ODN – sent out via email to all regional hospitals
Date placed on the websites (NWTS / PCC, SiC & LTV ODN) + MFT intranet	January 2025
Planned Review Date:	January 2028
Responsibility of:	Clinical lead North West & North Wales Paediatric Critical Care Network & NWTS guideline team
Minor amendment (if applicable) notified to:	**N/A**
Date notified:	**N/A**
EqIA Registration Number:	**2024-171**

1. Detail of Procedural Document

Management of Neonatal and Paediatric Hyperammonaemia

2. Equality Impact Assessment

Equality Impact Assessment Please record the decision whether the policy, service change or other key decision was assessed as relevant to the equality duty to: Eliminate discrimination and eliminate harassment Advance equality of opportunity Advance good relations and attitudes between people	
No concerns raised	
EqIA registration Number for RMCH:	**2024-171**

3. Consultation, Approval and Ratification Process

This guideline was developed with input from:

- North-West (England) and North Wales Paediatric Transport Service (NWTS).
- North-West (England) and North Wales Paediatric Critical Care Operational Delivery Network
- Representatives from the regional local hospitals within network above.
- Inborn Error of Metabolism consultants, RMCH

These guidelines were circulated amongst the North-West (England) and North Wales Paediatric Critical Care Operational Delivery Network for comments on the 11th April 2024 and 26th July 2024.
All comments received have been reviewed and appropriate amendments incorporated.

These guidelines were signed off by the PCC ODN guidelines committee on 27th January 2025.

For ratification process for network guidelines see appendix 1.

4. Disclaimer

These clinical guidelines represent the views of the North-West (England) and North Wales Paediatric Transport Service (NWTS) and the North-West (England) and North Wales Paediatric Critical Care Operational Delivery Network (PCCN). They have been produced after careful consideration of available evidence in conjunction with clinical expertise and experience.

It is intended that trusts within the Network will adopt this guideline and educational resource after review and ratification (including equality impact assessment) through their own clinical governance structures.

The guidance does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient.

Clinical advice is always available from NWTS on a case-by-case basis.

Please feel free to **contact NWTS (01925 853 550)** regarding these documents if there are any queries.

Management of Neonatal & Paediatric Hyperammonaemia

HYPERAMMONAEMIA = MEDICAL EMERGENCY

Prompt recognition, early treatment and transfer is vital for a good neurological outcome.

Symptoms & signs suggestive hyperammonaemia: lethargy/drowsiness, headache, encephalopathy, coma

Blood gas: metabolic acidosis or alkalosis

Stop feeds and start IV maintenance fluids 0.45% sodium chloride with 10% glucose.

CHECK ammonia (non-capillary sample; send on ice) AND blood gas + lactate + glucose + ketones

Assess & stabilise ABCDE

SEND: FBC, U&E, LFT, CRP, clotting, blood + urine cultures, Resp viral screen, blood acyl carnitines, plasma amino acids Urine amino & organic acids; urine toxicology. Hypoglycaemia screen IF blood glucose < 2.6 mmol/L (page 4)

TREAT for sepsis with broad spectrum antimicrobials including aciclovir (see sepsis guideline)

Ammonia ≥ 200 µmol/L

SEND repeat ammonia ASAP
2nd IV or IO access

ESCALATE management ASAP
Do **NOT** wait for results of repeat /2nd sample

Contact NWTS: 08000 84 83 82
Conference call with metabolic consultant

Ammonia = 100—200 µmol/L

SEND repeat ammonia ASAP
2nd IV or IO access

Ammonia > 200µmol/L

Ammonia = 100 - 200µmol/L

Discuss with metabolic team +/- NWTS if clinically necessary

Ammonia 50 - 100 µmol/L
Assess & stabilise ABCDE
Paediatric consultant review
D/W NWTS +/- metabolic consultant if appropriate

Consider repeating ammonia in 2-4 hours if suspect IEM

START infusions + fluid regime **within 30 minutes** of decision to treat (after d/w Metabolic consultant)
Sodium Phenylbutyrate, L-Arginine, Sodium Benzoate, L-Carnitine (dose/administration: pages 6 & 7)
Contact anaesthetic team/NICU consultant → **I&V to neuroprotect** patient

INTUBATE AND VENTILATE
Neuroprotective strategies: SpO_2 > 94%; ET CO_2 4-4.5; mean BP high normal (see page 8)

Consider carglumic acid (NWTS carry this)
After d/w metabolic consultant

SEND repeat ammonia level pre-departure & phone result to accepting centre

Ideally NWTS team to transfer to PICU.
BUT do NOT delay transfer. Aim => in tertiary centre within 6 hrs of identifying hyperammonaemia
NWTS unavailable? Neonate: d/w Connect NW on 0300 330 9299 ASAP
Infant/child/young person: time critical transfer by local team (use STOPP & d/w NWTS for advice on transfer)

Additional information available on BIMDG website *www.bimdg.org.uk* including drug calculator

HYPERAMMONAEMIA IS A MEDICAL EMERGENCY

Prompt recognition, early treatment and transfer of the child or young person is **vital to ensure a good neurological outcome**. Hyperammonaemia leads to direct neurological damage and outcome seems to be related to duration of hyperammonaemia and peak ammonia levels.

If a patient has an **ammonia level > 200 µmol/L, intravenous treatment must be started within 30 minutes** and urgent transfer to a tertiary paediatric centre organised ideally via NWTS.
If the ammonia > 400 µmol/L and is resistant to pharmacological treatment, aim to establish them on haemofiltration **within 6 hours** of identifying hyperammonaemia to achieve the best long-term outcome.

SYMPTOMS AND SIGNS OF HYPERAMMONAEMIA
can be subtle and varied, suspect & check ammonia if:

Neonate	Child or young person
Vomiting	Vomiting
Lethargy	Lethargy
Poor feeding	Ataxia
Encephalopathy	Seizures
Irritability	Encephalopathy
Pulmonary haemorrhage	Altered behaviour
Seizures	Signs of intoxication
Abnormal movements	Previous sudden death in family
Temperature instability	Unexplained metabolic acidosis
Low blood sugar	
Previous sudden death in family	
Unexplained metabolic acidosis OR alkalosis	

INVESTIGATIONS

- **Send an urgent ammonia - venous** (or arterial) **sample, NOT capillary.** Samples should be **free flowing** – capillary samples can give spuriously high results.
- **Send samples to the lab on ice and they should (ideally) be analysed within 20 minutes.**
- Phone and check lab staff are aware that the sample is being sent.
- NB Samples analysed after 20 minutes or not on ice and at room temperature will give a falsely elevated result. A normal result (sample analysed after > 20 mins) excludes hyperammonaemia.
- **If initial ammonia = 100-200 µmol/L, send repeat sample ASAP and d/w metabolic consultant.**
- **If initial ammonia > 200 µmol/L, send a repeat sample BUT ESCALATE management ASAP** without waiting for result of repeat /2nd sample **IE START** IV infusions and discuss with NWTS/Metabolic team
- **Site 2nd IV or IO access ASAP**
- **Investigations:** blood gas including lactate, glucose and ketones; FBC, U&E, LFT, clotting, plasma amino acids, acylcarnitines, and blood cultures.
- **Urine:** amino & organic (including orotic acid) acids (to be transferred with NWTS team to RMCH). Passing a urinary catheter allows rapid sample collection in critically sick patients.
- **Send: urine** for toxicology & culture, & **blood** for paracetamol, salicylates & alcohol levels.
- **Hypoglycaemia screen IF** symptomatic **blood glucose < 3 mmol/L** ie bloods for insulin, cortisol, c-peptide, thyroid function, growth hormone, B-hydroxybutyrate (ie ketones) if no point of care ketone testing available, free fatty acids and collect 1st urine passed. D/W endocrine team.
- **Watch potassium:** hypokalaemia is common, so monitoring essential. Consider adding potassium to maintenance fluids once urine output established and potassium level known.

ACUTE MANAGEMENT

- **Stop feeds + start IV maintenance fluids 0.45% sodium chloride with 10% glucose (ideally)** due to sodium content in both sodium butyrate & sodium phenylbutyrate infusions (if no alternative use 0.9% sodium chloride with 10% glucose).
- Maintenance with **10% glucose reduces:** nitrogen load, breakdown body protein, & ammonia production.
- Instructions on making intravenous fluids for metabolic patients are available on BIMDG website: *http://www.bimdg.org.uk/store/guidelines/intravenous_fluidsrev4_864191_09092016.pdf*
- **Contact NWTS on 08000 848382. NWTS will conference in the metabolic consultant on call at RMCH.**
- **Start broad spectrum antimicrobials including acyclovir** (see regional sepsis guideline)
- **Differential diagnosis raised ammonia** levels includes: sepsis, especially disseminated Herpes Simplex virus, low cardiac output states (eg congenital heart disease, cardiomyopathy), and liver failure
- **START metabolic drug infusions within 30 minutes as directed by metabolic team (see appendix).**
- **Ammonia can rise exponentially in a decompensated metabolic disorder.**
- **ALL HOSPITALS** in North West (England) and North Wales region **must maintain** a supply of ***intravenous* sodium phenylbutyrate, sodium benzoate, L-arginine (arginine hydrochloride) and L-carnitine.**
- **Delays > 30 minutes starting treatment increases risk of neuronal injury and worse neurological outcome.** Requesting drugs acutely from RMCH or Alder Hey leads to a huge delay starting treatment and leads to a worse neurological outcome due to prolonged hyperammonaemia.
- **Contact local anaesthetic team +/- neonatal team to intubate and ventilate ASAP** for transfer even if no respiratory difficulty. Ventilation reduces metabolic demands on the patient and so reduces ammonia production.
- **Ammonia is a potent neurotoxin**, causing cerebral oedema / raised intracranial pressure which increases the risk of cerebral herniation or coning. Once intubated and ventilated use neuroprotective strategy.
- **Neuroprotective strategies once I&V** aim for: SpO_2 ≥ 94%; ET CO_2 4-4.5; mean BP high normal **(resources page 8)**
- **Avoid using propofol bolus or infusion:** in IEM or mitochondrial disorders as these patients are at increased risk of propofol infusion syndrome especially with use ≥ 48 hours. **Use morphine/midazolam infusions for sedation**.
- Critically sick patients are expected to have blood glucose > 3 mmol/L due to stress response.
- **TREAT hypoglycaemia IF blood glucose < 3 mmol/L ie** give 10% glucose 3 mL/kg and start maintenance fluids containing 10% glucose (as above). Recheck blood glucose after bolus and repeat when on maintenance fluids.
- **If shocked** give 10 mL/kg fluid bolus using balanced crystalloid (Plasmalyte 148 or Hartmann's solution) immediately after glucose bolus. Re-assess and repeat fluid bolus +/- start inotropes as per regional guideline.
- Hyperglycaemia may be a problem. If blood glucose exceeds 14 mmol/L and glycosuria is present, do not reduce glucose concentration in fluids, but consider starting an insulin infusion (as per diabetic guidelines).

PRE-TRANSFER:

- **Carglumic acid may be given** on advice from Metabolic consultant on call. It is not stocked by most referring hospitals so will be brought by the transport team. It is given as a single enteral dose (via NGT).
- **SEND a repeat / 3rd ammonia sample pre-departure from the local referring hospital.** Contact NWTS with the result ASAP as this determines if the patient needs haemofiltration soon after arrival on PICU.

TRANSFER:

- **Ideally transfer patient to PICU at RMCH if possible for easier access to the on-site metabolic team**
- **Transfer to PICU** should be undertaken **within 6 hours of identification of hyperammonaemia**.
- **If NWTS are unable to undertake the transfer within 6 hours of identification of hyperammonaemia**:
  - If a neonate NWTS will discuss with Connect NW (regional neonatal transport team on 0300 330 9299) to organise transfer to PICU for definitive treatment (ie haemofiltration).
  - If infant/child/young person or Connect NW not available, the local team will need to undertake an urgent transfer to PICU for definitive treatment (ie haemofiltration). D/W NWTS for stabilisation and transfer advice and use STOPP document (***www.nwts.nhs.uk/clinicalguidelines***).

DRUGS FOR METABOLIC DECOMPENSATION for patients less than 10 kg

SODIUM BENZOATE

Loading Dose: **250mg/kg over 90minutes**
Rate: **5mL/kg over 90minutes** (use concentration below)
Maintenance Dose: **250mg/kg per DAY by continuous infusion**
Rate: **0.2mL/kg/hr** (use concentration below)

PREPARATION using Sodium Benzoate **1g/5mL (20%)** solution: Draw up 12.5mL and make up to 50mL with 10%glucose. Mix well (tip up/down approximately 20 times). Final concentration 50mg/1mL (5%).

Sodium content of daily maintenance dose 3.5 mmol/kg.

SODIUM PHENYLBUTYRATE

Loading Dose: **250mg/kg over 90minutes**
Rate: **5mL/kg over 90minutes** (use concentration below)
Maintenance Dose: **250mg/kg/day by continuous infusion**
Rate: **0.2mL/kg/hr** (use concentration below)

Preparation using Sodium Phenylbutyrate **1g/5mL (20%)** solution: Draw up 12.5mL and make up to 50mL with 10% glucose. Mix well. Final concentration 50mg/1mL (5%).

Sodium content of daily maintenance dose 2.8 mmol/kg.

L-ARGININE

Loading Dose: **150mg/kg over 90 minutes**
Rate: **3mL/kg over 90 minutes** (use concentration below)
NB For some patients the metabolic consultant may advise 300mg/kg over 90 minutes
ie Rate: 6 mL/kg over 90 minutes (using concentration below)
Maintenance Dose: **150-300mg/kg per DAY by continuous infusion**
Rate: **0.12 – 0.26mL/kg/hour** (use concentration below)

Preparation using L-Arginine **100mg/mL (10%)** solution: Draw up 25mL and make up to 50mL with 10% glucose. Mix well. Final concentration 50mg/1mL (5%). This is maximum concentration that can be used via peripheral lines.

CAUTION: other L-Arginine vial concentrations are available—if used check calculations very carefully.

CARGLUMIC ACID

NWTS WILL BRING CARGLUMIC ACID WITH THEM
Loading Dose: **250mg/kg as a single ENTERAL dose**

Preparation: Disperse 200 mg tablet in 5mL of water to give 40 mg per mL solution. Shake gently. Draw up the appropriate volume & administer immediately via nasogastric tube (NGT). Flush NGT with additional water to clear.

L-CARNITINE

Dose: **25mg/kg FOUR times a day (Max 3 grams / day)**
Administration: **Give as a bolus over 2-3 minutes.**
Occasionally the metabolic team will request this to be run as an infusion.
The dose for this is on BIMDG website *www.bimdg.org.uk*

WARNING: Should NOT be used if any evidence of cardiomyopathy, any cardiac arrythmias or if a long chain fatty acid oxidation disorder is suspected — always discuss with Metabolic consultant first.

PREPARATION: Use the L-Carnitine 1g/5mL (20%) solution for injection. The bolus can be administered undiluted, however **always** consider **diluting** as high osmolality and high risk of extravasation injury. For infusion, dilution is recommended up to 50mg/mL with 10% glucose OR sodium chloride 0.9%. See Medusa IV guide.

Doses will vary with different metabolic disorders.
Always follow the guidance on doses given by the Metabolic consultant on call at RMCH
ALL infusions can be administered via a PERIPHERAL line
Infusions are COMPATIBLE WITH EACH OTHER and can run ON THE SAME LINE (see page 7)
Check Medusa IV guide for compatibility for any other fluids used.

DRUGS FOR METABOLIC DECOMPENSATION: Patients OVER 10 kg

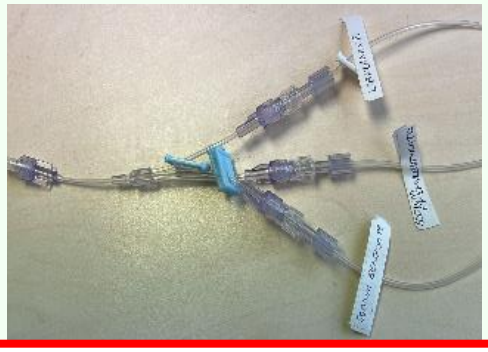
Doses are unchanged but for ease of administration, the following method of making up the infusions is recommended

Sodium Benzoate 2.5% and Sodium Phenylbutyrate 2.5% (in 500 mL 10% glucose) combination For loading dose and infusion

Using products available: Sodium Benzoate 1g/5mL (20%) ampoules
Sodium Phenylbutyrate 1g/5mL (20%) ampoules
500mL bag of 10% glucose.
Preparation: Draw out and discard 125mL from 500mL bag of 10% glucose.
Draw up 62.5mL of 20% Sodium Benzoate **AND** 62.5mL of 20% Sodium Phenylbutyrate.
Add both drugs to 10% glucose 500 mL bag.
Mix well (tip up/down min. 10 times). Final concentration = 2.5g/100mL (2.5%).

Rate of infusion to deliver Loading Dose: **10mL/kg over 90 minutes.**
This provides 250mg/kg of sodium benzoate and 250mg/kg of sodium phenylbutyrate.
Rate of infusion to deliver Maintenance Dose: **0.42mL/kg/hour**
This provides 250mg/kg/DAY of sodium benzoate and 250mg/kg/DAY of sodium phenylbutyrate.

L-Arginine As per infants under 10 kg



Please label each syringe or fluid bag with drug concentration.

Sodium Benzoate, Sodium Phenylbutyrate and L-Arginine infusions can all run together via a single PVL.

These drugs do not need 3-way taps in the lines.

North-West (England) & North Wales region Paediatric Critical Care ODN STRONGLY RECOMMENDS that EVERY HOSPITAL can access ALL the DRUGS required to treat metabolic decompensation WITHIN 30 minutes.

This includes:

1. Sodium benzoate 1g in 5mL ampoules (20 ampoules)
2. Sodium phenylbutyrate 1g in 5mL ampoules (20 ampoules)
3. L-Arginine 10% 200mL vials (2 vials). Other strengths are acceptable.
4. L-Carnitine 1g in 5mL ampoules (5 ampoules)

RECOMMENDATIONS FOR PHARMACY DEPARTMENTS: these drugs are time critical and appropriate stock must always be available for all age groups 24/7.

RECOMMENDATIONS FOR REGIONAL TRANSPORT TEAMS:
1. Local referring hospitals are not required to stock carglumic acid, but this can have a dramatic effect at reducing ammonia levels in some patients.
2. All transport teams should stock a small amount of carglumic acid so that when they reach the patient a dose can be given if advised by the metabolic team.

RESOURCES: including quick reference guide for National PEWS

TARGETS for managing patient with hyperammonaemia:

ALL AGES	SpO_2 ≥ 94%	ET CO_2: 4-4.5 kPa	Glucose: ≥ 3 mmol/L
AGE	**TARGET MEAN BP**	**AGE**	**TARGET MEAN BP**
0-11 Months	**45-55**	5-12 Years	**60**
1-4 Years	**55-60**	>13 Years	**60-65**

NPEWS: Respiratory Rate (Score up to 4)							
Score	**4**	**2**	**1**	**0**	**1**	**2**	**4**
0-11 months	≤ 10	11-20	21-20	31-49	50-59	60-69	≥70
1-4 years	≤ 10	11-20		21-39	40-49	50-59	≥ 60
5-12 years	≤ 10	11-15	16-20	21-24	25-39	40-49	≥ 50
>13 years	≤ 10		11-15	16-24	25-29	30-39	≥ 40

ALL AGES Score	NPEWS: Respiratory Distress (Score up to 4)
0 = none	None
1 = mild	Nasal flaring, subcostal recession
2 = moderate	Tracheal tug, intercostal recession, inspiratory or expiratory noises
4 = severe	Supraclavicular recession, grunting, exhaustion, impending respiratory arrest

ALL AGES Score	NPEWS: Oxygen Saturations (Score up to 4)
0	95-100%
2	92-94%
4	≤ 91%

ALL AGES Score	NPEWS: Oxygen Requirement (Score up to 4) - ALL AGES
0	Room Air
2	0.01 up to 4 litres/min
4	4 or more litres/min **NB** High flow humidified NC oxygen, NIV CPAP or BiPAP score 4 (irrespective of oxygen requirement)

NPEWS: Heart Rate (Score up to 4)							
Score	**4**	**2**	**1**	**0**	**1**	**2**	**4**
0-11 Months	≤ 80	81-90	91-110	111-149	150-169	170-179	≥ 180
1-4 Years	≤ 60	61-70	71-90	91-139	140-149	150-169	≥ 170
5-12 Years	≤ 60	61-70	71-80	80-119	120-139	140-159	≥ 160
>13 Years	≤ 50	51-60	61-70	71-99	100-119	120-129	≥ 130

NPEWS: Blood Pressure Systolic (Score up to 4)							
Score	**4**	**2**	**1**	**0**	**1**	**2**	**4**
0-11 Months	≤ 50	51-60	61-70	71-89	90-99	100-109	≥ 110
1-4 Years	≤ 50	51-60	61-80	81-99	100-119	120-129	≥ 130
5-12 Years	≤ 70	71-80	81-90	91-109	110-119	120-129	≥ 130
>13 Years	≤ 80	81-90	91-100	101-119	120-129	130-139	≥ 140

NPEWS: Capillary Refill Time (CRT) (Score up to 2)							
Score	**4**	**2**	**1**	**0**	**1**	**2**	**4**
All Ages		≥ 3 secs		<3 secs		≥ 3	

CHECK IF YOUR PATIENT HAS ANY ADDITIONAL RISK FACTORS (NPEWS)		
RISK FACTOR	**THINK!**	
☐ Baseline vital signs outside normal reference ranges	Always score relevant PEWS value even if this is normal for the patient eg cyanotic heart disease	Vital sign: Patient's normal value:
☐ Tracheostomy / Airway Risk	Do you need additional help in an airway emergency? Needs review by local anaesthetics & ENT teams. Consider d/w NWTS early	
☐ Invasive/Non-invasive ventilation/high flow	Check oxygen requirement on additional respiratory support. Remember High Flow/BiPAP & CPAP score max 4 on oxygen delivery	
☐ Neutropenic/immunocompromised	Sepsis recognition & escalation has a lower threshold	
☐ <40 weeks corrected gestational age	Sepsis recognition & escalation has a lower threshold (beware hypothermia)	
☐ Neurological condition (ie meningitis, seizures)	Remember: check pupil response if anything other than ALERT on AVPU	
☐ Neurodiversity or Learning Disability	Be aware of the range of responses to pain & physiological changes	

NPEWS ESCALATION LEVEL	ACTIONS	MEDICAL REVIEW	OBSERVATIONS / PLAN
E0 – no concerns **Score = 0**	None	Not required	Continue current observations
E1 – Increased monitoring **Score = 1- 4**	Inform Nurse-in-Charge Consider medical review (ST3+ or equivalent) Ensure feedback to parents	As required Discuss with Nurse-in-Charge whether medical review required	Reassess within 60 mins & document ongoing plan
E2 – Needs clinical review (within 30 mins) **Score = 5-8**	Review by Nurse-in-Charge Ensure feedback to parents	Within 30 mins Review by ST3+ or equivalent Discuss stabilisation plan with consultant	Reassess within 30 mins & document ongoing plan Continuous SpO_2 monitoring
E3 – Needs rapid review (within 15 mins) **Score = 9-12**	Immediate review by Nurse-in-charge Discuss medical plan with consultant Senior feedback to parents	Within 15 mins Alert to ST3+ or equivalent Stabilisation plan to be agreed after review by consultant Consider NWTS referral after consultant review	Reassess every 30 mins Continuous monitoring SpO_2, RR, & ECG Record full GCS if change in AVPU
E4 – Needs emergency review (immediate) **Score > 12**	Immediate review by Nurse-in-Charge Consider immediate 2222 call for immediate emergency medical response Inform paeds consultant Senior feedback to parents	Immediate Alert to ST3+ or equivalent Consultant review ASAP Anaesthetic review Consider NWTS referral after appropriate initial interventions	Reassess every 15 mins Continuous SpO_2, ECG, & RR Record full GCS if change in AVPU

NB Escalation levels can also be selected and triggered if parent or carer expresses concern that their child needs increased monitoring +/- clinical review despite PEWS, OR parent or nursing gut instinct irrespective of score.

Medical Plan for Stabilisation:

Structured plan must be documented including:

1. Specific actions to be taken
2. Expected outcome
3. Outcome deadline / in timeframe
4. Escalation if outcome not met by deadline / in timeframe

DRUG AND INFUSION GUIDES

BIMDG IV drugs calculator (for all metabolic drugs):
http://www.bimdg.org.uk/store/guidelines/UCDcalculator_for_UNDIAGNOSED_v6-1- 289867-04-12-2013_288879_16042017.xls

Instructions on making intravenous fluids for metabolic patients are available on BIMDG website.
http://www.bimdg.org.uk/store/guidelines/intravenous_fluidsrev4_864191_09092016.pdf

NWTS emergency drugs guide via NWTS website home page - for intubation drugs / sedation regime / inotropes etc ***https://www.nwts.nhs.uk***

FOR OTHER DRUG DOSES: **British National Formulary for Children**

GUIDELINES FOR < 16 YEARS: ***www.nwts.nhs.uk/clinicalguidelines***

STOPP tool = Safe Transfer of Paediatric Patients which includes risk assessment prior to transfer, and checklists
NWTS LocSIPPS includes sizes of ETT, suction, NGT, CVL & arterial lines and checklist for paediatric intubation
Guidelines include: intubation and difficult airway, sepsis including inotropes, insertion of intraosseous line, collapsed neonate or infant, management of under 16 years outside PCC level 3 unit, and transfer

EDUCATION: ***www.nwts.nhs.uk/education-website***
Includes recordings of NWTS education eg time critical transfers, sepsis, airway management etc
Login details for NWTS education site are available from your nursing, AHP and medical paediatric critical care operational delivery network links
OR via email: info@nwts.nhs.uk

CONTACT NUMBERS:

NWTS (North-West (England) & North Wales Paediatric Transport Service): **Referrals 08000 84 83 82**
General enquiries: 01925 853 550
Regional Paediatric Intensive Care Unit Alder Hey Childrens Hospital: 0151 252 5241
Regional Paediatric Intensive Care Unit Royal Manchester Childrens Hospital: 0161 701 8000
Regional Inborn Errors of Metabolism Consultant on call via switchboard at RMCH: 0161 276 1234

Consulted parties:
Inborn Errors of Metabolism Consultant Team, RMCH
North-West (England) & North Wales Paediatric Transport Service (NWTS)
North-West (England) and North Wales Paediatric Critical Care Operational Delivery Network
PICU teams at both Royal Manchester Children's Hospital & Alder Hey Children's Hospital
North-West (England) Neonatal Operational Delivery Network

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Next Review Due: January 2028

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