BNSSG Paediatric Shared Care Guidance

Please complete all sections

Section 1: Heading

Drug	Sodium benzoate
Amber ***three months***	
Indication	Hyperammonaemia Non-ketotic hyperglycinaemia (NKH) Guanidinoacetate methyltransferase (GAMT) deficiency
Speciality / Department	Department for Paediatric Metabolic Diseases
Trust(s)	University Hospitals Bristol NHS Foundation Trust

Section 2: Treatment Schedule

Usual dose and frequency of administration *(Please indicate if this is licensed or unlicensed for this age group and any relevant dosing information)*	Hyperammonaemia/Urea cycle defects: 50 – 100 mg/kg TDS - QDS (250 mg/kg/day). Maximum of 12 grams a day *All doses are maintenance doses, not IV doses for acute hyperammonaemia. Acute hyperammonaemia requires urgent medical attention from a hospital.* Non-ketotic hyperglycinaemia (NKH): 250 – 750 mg/kg/day in 4 divided doses, Maximum of 12 grams a day. Guanidinoacetate methyltransferase (GAMT) deficiency:100 mg/kg/day divided doses.
Route and preferred formulation *(Please indicate licensed or unlicensed preparation)*	Oral liquid: 500 mg/5mL (unlicensed product such as “Amzoate” from Special Products) Oral tablets: 500 mg (unlicensed product such as “Amzoate” from Special Products) Must be of a pharmaceutical grade, such as products above used by Bristol Children’s Hospital as pharmaceutical quality of the product has been assessed by Pharmacy QC department, UHBristol.
Duration of treatment	Life-long

Section 3: Monitoring

Please give details of any tests that are required before or during treatment, including frequency, responsibilities (please state whether they will be undertaken in primary or secondary care), cause for adjustment and when it is required to refer back to the specialist)

Baseline tests to be done by secondary care

Ammonia, Urea & Electrolytes (U&Es), Liver Function Test (LFTs), Full Blood Count (FBC), Plasma amino acid profile

Subsequent tests - where appropriate *(Please indicate who takes responsibility for taking bloods and interpreting results. If the drug is dosed by weight please also indicate intended frequency of weight monitoring/dose adjustment)*

Test	Frequency	Who by	Action/management
U&Es	4-6 monthly with outpatient clinic appointment	Secondary Care, Department for Paediatric Metabolic Diseases	All action and management, including dose to be adjusted as per weight etc to be directed by Secondary Care, Department for Paediatric Metabolic Diseases.
LFTs	4-6 monthly with outpatient clinic appointment		
FBCs	4-6 monthly with outpatient clinic appointment		
Ammonia	4-6 monthly with outpatient clinic appointment		
Plasma amino acid profile	4-6 monthly with outpatient clinic appointment		

Frequency of ongoing follow up by secondary care *(Please indicate how often child will continue to be seen by secondary care i.e. at least every 6 months)*	Every 4 to 6 months as above.

Section 4: Side Effects

Please list only the most pertinent side effects and management. Please provide guidance on when the GP should refer back to the specialist. For everything else, please see BNFc or SPC.

Side effects and management

Side effect	Frequency/severity	Action/management
Hypernatraemia	Common	Contact Secondary care
Anorexia	Frequency not known	Contact Secondary care
Coma	Frequency not known	Contact Secondary care
Irritability	Frequency not known	Contact Secondary care
Nausea	Common	Take with food to reduce nausea and vomiting and contact secondary care
Vomiting	Common	
Reflux	Uncommon	GI side effects may be reduced by giving in smaller divided doses and consider gastro protection with PPI, and contact secondary care.
Hypokalaemia	Common	Contact secondary care for potassium

	supplementation
Referral back to specialist	Any medical or clinical concern

Section 5: Other Issues
(e.g. Drug Interactions, Contra-indications, Cautions, Special Recommendations)

Please list only the most pertinent and the action for GP to take (For full list please see BNFc or SPC)

Issues	Caution in patients with oedema and sodium retention Caution in patients with congestive heart failure Caution in neonates (risk of kernicterus with neonatal hyperbilirubinemia)
Reminder to ask patient about specific problems	N/A

Section 6: Advice to the patient

Advice for prescribing clinician to inform patient

Take with food/meals

Section 7: Generic principles of shared care for SECONDARY CARE

Core responsibilities

1. Initiating treatment and prescribing for the length of time specified in **section 1.**
2. Undertaking the clinical assessment and monitoring for the length of time specified in **section 1** and thereafter undertaking any ongoing monitoring as detailed in **section 3.**
3. Communicate details of the above in 1 and 2 to GP within the first month of treatment. This information should be transferred in a timely manner.
4. Refer patients to GP and provide information of further action where appropriate e.g. if blood test is due.
5. To provide advice to primary care when appropriate.
6. Review in frequency specified in **section 3** and adjust dose for child's age/body weight as appropriate.
7. Review concurrent medications for potential interaction prior to initiation of drug specified in **section 1.**
8. Stopping treatment where appropriate or providing advice on when to stop.
9. Reporting adverse events to the MHRA.
10. Reminder to ask patients about particular problems see **section 5.**

Section 8: Generic principles of shared care for PRIMARY CARE

Core responsibilities

1. Responsible for taking over prescribing after the length of time specified in **section 1.**
2. Responsible for any clinical assessment and monitoring if detailed in **section 3** after the length of time specified in **section 1.**
3. Review of any new concurrent medications for potential interactions.
4. Reporting adverse events to the MHRA.
5. Refer for advice to specialist where appropriate.
6. Reminder to ask patients about particular problems see **section 5.**

Section 9: Contact Details

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Section 10: Document Details

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Section 11: Collaboration

Specialists in any one discipline are encouraged to collaborate across the health community in preparing shared care guidance. Please give details

1. Click here to enter details

Section 12: References

Please list references

1. Saudubray *et. al.*2016. Inborn Metabolic Diseases: Diagnosis & Treatment. 6th Edition. ISBN978-6-662-49769-2
2. British National Formulary for Children. Accessed online from: https://bnfc.nice.org.uk/ on 26/06/2018
3. Evelina Formulary. Accessed online from: http://cms.ubqo.com/public/d2595446-ce3c-47ff-9dcc-63167d9f4b80 on 26/06/2018
4. Hoffmann, G., Zschocke, J., Vademecum Metabolicum: Diagnosis & Treatment of Inborn Errors of Metabolism. E-book can be accessed online from: http://www.vademetab.org/
5. Stocker-Ipsiroglu S., Van Karnebeek, C., Longo, N., *et. al.,* 2014. Guanidinoacetate methyltransferase (GAMT) deficiency: Outcomes in 48 individuals and recommendations for diagnosis, treatement and monitoring. *Molecular Genetics and Metabolism*; 111 : 16- 25