

BNSSG Paediatric Shared Care Guidance

Please complete all sections

Section 1: Heading

Drug	L-Arginine (Arginine)
Amber <i>three months</i>	
Indication	<p>Urea cycle disorders of which include:</p> <ul style="list-style-type: none"> - Ornithine Transcarbamylase (OTC) deficiency - Carbamylphosphate synthase I (CPSI) Deficiency - Citrullinaemia type I - Arginosuccinic aciduria (ASA) - Citrullinaemia type II (Citrin deficiency) <p>Mitochondrial encephalomyopathy, lactic acidosis and stroke like episodes (MELAS)</p>
Speciality / Department	Department for Paediatric Metabolic Diseases
Trust(s)	University Hospitals Bristol NHS Foundation Trust
	North Bristol NHS Trust

Section 2: Treatment Schedule

Usual dose and frequency of administration (Please indicate if this is licensed or unlicensed for this age group)	<p>All doses are maintenance doses, not IV doses for acute hyperammoniaemia. Acute hyperammoniaemia requires urgent medical attention from a hospital.</p> <p>OTC and CPS: 100 – 250 mg/kg/day in 3 – 4 divided doses, maximum of 6 grams a day. Citrullinaemia type I & ASA: up to 300 mg/kg/day in 3 – 4 divided doses, maximum of 6 grams a day. Citrin deficiency: up to 15 grams a day in adolescents MELAS(oral maintenance dose): 150 – 300 mg/kg/day in 3 divided doses</p>
Route and preferred formulation (Please indicate licensed or unlicensed preparation)	<p>Oral liquid: 100 mg/mL (unlicensed from Special Products) Oral Capsules: 500 mg (unlicensed from Martindale Pharma) Oral tablets: 1 gram (unlicensed/food supplement from Lamberts)</p> <p>Must be of a pharmaceutical grade, such as products above used by Bristol Children’s Hospital.</p>

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Relevant dosing information	N/A
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			
Urea & Electrolytes (U&Es), Liver Function Test (LFTs), Full Blood Count (FBC), Bicarbonate, Chloride, plasma pH, Plasma amino acid profile, Blood Pressure (BP)			
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	As per clinic appointment	Secondary Care, Department for Paediatric Metabolic Diseases	All action and management to be directed by Secondary Care, Department for Paediatric Metabolic Diseases
LFTs	As per clinic appointment		
FBCs	As per clinic appointment		
Bicarbonate	As per clinic appointment		
Plasma amino acid profile	As per clinic appointment		
Blood Pressure	As per clinic appointment	Secondary Care, Department for Paediatric Metabolic Diseases	All action and management to be directed by Secondary Care, Department for Paediatric Metabolic Diseases

Section 4: Side Effects

Please list the most common side effects and management. Please provide guidance on when the GP should refer back to the specialist.

Side effects and management	Side effect	Frequency/severity	Action/management
	Flushing	Frequency not known	Secondary Care, Department for Paediatric Metabolic Diseases
	Headache	Frequency not known	
	Hyperchloraemic metabolic acidosis	Frequency not known	
	Hypotension	Frequency not known	
	Nausea	Frequency not known	
	Vomiting	Frequency not known	
	Numbness	Frequency not known	
Referral back to specialist	Any medical or clinical concern		

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Section 5: Drug Interactions

Please list clinically significant drug interactions ([eMC link](#) please click here)

Significant Drug Interactions	Mono and di- nitrates. (Arginine is a pre-cursor of nitric oxide and can therefore cause hypotension and should not be given with nitrates)
Reminder to ask patient about specific problems	N/A

Section 6: Contra-indications, Cautions and Special Recommendations

Please list

Arginase deficiency – contraindication
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Section 7: Advice to the patient

Advice for prescribing clinician to inform patient

Take with food

Section 8: Responsibilities for Secondary Care

Core responsibilities <ol style="list-style-type: none">1. Initiating treatment and prescribing for the first three months2. Undertaking the clinical assessment and monitoring for the first three months.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. blood test is due.5. To provide advice to primary care when appropriate.6. Review concurrent medications for potential interaction prior to initiation of L- arginine (Arginine).7. Stopping treatment where appropriate or providing advice on when to stop.8. Reporting adverse events to the MHRA.9. Reminder to ask patients about particular problems see section 5.
Other specific to drug
N/A

Section 9: Responsibilities for Primary Care

Core responsibilities <ol style="list-style-type: none">1. Responsible for taking over prescribing after the first three months2. Responsible for the clinical assessment and monitoring after the first three months3. 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.
Other specific to drug

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N/A

Section 10: Contact Details

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Section 11: Document Details

Date prepared	26/06/2018
Prepared by	Will Batten, Paediatric Specialist Pharmacist
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Section 12: Collaboration

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

N/A

Section 13: References

Please list references

<ol style="list-style-type: none">1. Saudubray <i>et. al.</i> 2016. Inborn Metabolic Diseases: Diagnosis & Treatment. 6th Edition. ISBN978-6-662-49769-22. British National Formulary for Children. Accessed online from: https://bnfc.nice.org.uk/ on 26/06/20183. Evelina Formulary. Accessed online from: http://cms.ubqo.com/public/d2595446-ce3c-47ff-9dcc-63167d9f4b80 on 26/06/20184. Hoffmann, G., Zschocke, J., Vademecum Metabolicum: Diagnosis & Treatment of Inborn

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Errors of Metabolism. E-book can be accessed online from: <http://www.vademeta.org/>