

BNSSG Shared Care Guidance

Please complete all sections

Section 1: Heading

Drug	Co-enzyme Q10 (ubiquinone/ubidecaranone)
Amber <i>three months</i>	
Indication	Inborn errors of Q10 synthesis Mitochondrial cytopathies
Speciality / Department	Paediatric Metabolic Diseases
Trust(s)	University Hospitals Bristol NHS Foundation Trust

Section 2: Treatment Schedule

Usual dose and frequency of administration	Normal dose for mitochondrial disorders is 100 – 300 mg a day. Starting doses normally begin at 10 mg/kg and maximum doses of 30 mg/kg/day. Usual adult dose is 1 - 4 gram a day so routine to cap dose for children at adult dose. Normally, daily dose is given in 1 to 2 divided doses i.e. ONCE or TWICE a day
Route and formulation	Routes: oral, via nasogastric and PEG tubes Formulations: 30mg and 100mg capsules available from PharmaNord. Administration instructions: Capsules to be pierced and capsule contents to be given orally, via nasogastric tube or PEG tubes. Can be given with a small amount of food/liquid.
Duration of treatment	Lifelong until no longer appropriate

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 - where appropriate
Muscle and skin biopsies performed at diagnosis (or soon after suspected diagnosis) to be undertaken in secondary care. Usually happens before transfer to primary care. Co-enzyme Q10 administration withheld 7 days prior to biopsy.

BNSSG Shared Care Guidance

Subsequent tests - where appropriate
1. Liver Function Test (will be taken and interpreted by secondary care)

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	Common or very common: diarrhoea, heartburn, nausea Rare: agitation/alertness/wakefulness, dizziness, headache, irritability
Referral back to specialist	Any of above. Wakefulness can sometimes be alleviated by taking a larger portion of daily dose in the morning. GI upset (cramps, aches, nausea and diarrhoea) can be alleviated by taking co-enzyme Q10 with food.

Section 5: Drug Interactions

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

Significant Drug Interactions	Ubidecarenone may reduce or enhance the effect of warfarin.
Reminder to ask patient about specific problems	Nil

Section 6: Contra-indications, Cautions and Special Recommendations

Please list

1. Caution: May reduce insulin requirement in diabetes mellitus 2. Reduce dose in moderate and severe hepatic impairment

Section 7: Advice to the patient

Advice for prescribing clinician to inform patient

1. Advice will provided by hospital initiating treatment. 2. Further reputable patient friendly information can be found at: http://www.medicinesforchildren.org.uk/ubidecarenone-mitochondrial-disease
--

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.
--

BNSSG Shared Care Guidance

5. To provide advice to primary care when appropriate.
6. Review concurrent medications for potential interaction prior to initiation of any medication.
Paracetamol and ibuprofen are okay to give alongside co-enzyme Q10.
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

All therapeutic drug monitoring to be undertaken by secondary care

Section 9: Responsibilities for Primary Care

Core responsibilities

1. Responsible for taking over prescribing after the first three months
2. Responsible for the clinical assessment and monitoring after the first three months
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.

Other specific to drug

Bristol Childrens Hospital recommend supplying patient with PharmaNord brand of co-enzyme Q10.

Section 10: Contact Details

Name	Organisation	Telephone Number	E mail address
Dr. Germaine Pierre	Univerisity Hospitals Bristol NHS Foundation Trust	0117 342 1694	Germaine.Pierre@UHBristol.nhs.uk
Dr. Effie Chronopoulou	University Hospitals Bristol NHS Foundation Trust	0117 342 1694	Efstathia.Chronopoulou@UHBristol.nhs.uk
Will Batten	Univeristy Hopsitals Bristol Foundation Trust	0117 342 7042	William.Batten@UHBristol.nhs.uk

Section 11: Document Details

Date prepared	17/08/2017
Prepared by	Will Batten
Date approved by PJFG	November 2017

BNSSG Shared Care Guidance

Date of review	November 2019
Document Identification: Version	V1.2

Section 12: 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 13: References

Please list references

1. Saudebray, 2011, Inborn Metabolic Diseases: Diagnosis and Treatment, 5th Edition, Springer.
2. British National Formulary for Children. Accessed online on 26/04/2016 via NHS Evidence.
3. Guys and St Thomas', Kings college and University Lewisham Hospitals, Paediatric Formulary, 9th Edition, 2012
4. Zschocke and Hoffmann, 2004, Vademecum metabolicum: Manual of metabolic paediatrics, 3rd Edition, Milupa Schattauer
5. Parikh et al, 2009, A Modern Approach to the Treatment of Mitochondrial Disease, *Current Treatment*