BSW APC: BaNES, Swindon & Wiltshire (BSW) CCG, Avon & Wiltshire Mental Health Partnership NHS Trust (AWP), Royal United Hospitals Bath NHS Foundation Trust, Great Western Hospitals NHS Foundation Trust, Salisbury NHS Foundation Trust, Virgin Care, Swindon Community Health Services, Wiltshire Health & Care

SHARED CARE AGREEMENT

OFF-LABEL Co-enzyme Q10 (ubiquinone/ubidecarenone) for Inborn errors of Q10 synthesis; Mitochondrial cytopathies – Children

Amber TLS – 3 Months

Principles of Shared Care

Shared care agreements provide a framework for the seamless transfer of care from a hospital or specialist service setting to general practice, where this is appropriate and in the patient's best interest. When a specialist considers a patient's condition to be stable or predictable, they may seek the agreement of the GP (or other primary care prescriber) concerned and the patient to share their care.

Patients and/or carers must be centrally involved in any decision-making process. They should be supported by good quality information that helps them to both come to an informed decision about engagement in a shared care arrangement and sets out the practical arrangements for ongoing supplies of medicines.

The existence of a shared care agreement does not necessarily mean that the GP has to agree to and accept clinical and legal responsibility for prescribing; they should only do so if they feel clinically confident in managing that condition. Clinical responsibility for prescribing is held by the person signing the prescription, who must also ensure adequate monitoring.

Responsibilities of Secondary Care Specialist

- Initiate treatment and prescribe for the length of time agreed (3 months) – this should be a sufficient amount of time to allow optimisation of treatment and demonstrate that the patient's response is consistent.
- Discuss the benefits and side effects of treatment with the patient.
- Review concurrent medications for potential interactions prior to initiation.
- Undertake the clinical assessment and relevant monitoring at baseline and during the initiation period.
- Communicate details of treatment to GP (in writing or via secure email) within the first month of treatment and ask the GP whether he or she is willing to participate in shared care.
- Discuss shared care arrangements with the patient/carer, obtain their consent and explain their responsibilities.
- Review the patient's condition and monitor response to treatment regularly where indicated.
- Inform the GP after each clinic attendance if there is any change to treatment or monitoring.
- Supply GP with clinic letter or discharge summary within 14 days of an outpatient review or inpatient admission, and inform GP if patient does not attend scheduled clinic appointments.
- Ensure that clear arrangements exist for GPs to obtain advice and support.
- Report adverse events to the MHRA.
- Stop treatment where appropriate or provide GP with advice on when to stop.

Responsibilities of GP/Primary Care Prescriber

- Reply to the request as soon as practicable if they are **unable** to support shared care (in writing or via secure email).
- Prescribe medicine at the dose recommended after the initiation period.
- Undertake ongoing clinical assessment and relevant monitoring following initiation period.
- Review any new concurrent medications for potential interactions.
- Refer promptly to specialist when any loss of clinical efficacy is suspected (e.g. worsening of disease-related symptoms, new symptoms suggestive of disease recurrence or progression) or intolerance to therapy occurs.
- Report to and seek advice from the specialist on any aspect of patient care that is of concern and may affect treatment.
- Report adverse events to the specialist and MHRA.
- Stop treatment on the advice of the specialist.

Responsibilities of Patient/Carer

- Report to the specialist or GP if he or she does not have a clear understanding of the treatment.
- Share any concerns in relation to treatment with medicine.
- Report any adverse effects to the specialist or GP whilst taking the medicine.
- Attend appointments for clinical review and monitoring.

<table>
<tr><td>1. Summary of condition and treatment aims
Include links to relevant clinical guidelines e.g. NICE</td><td colspan="2">Mitochondrial Respiratory Chain Disorders– heterogenous group of metabolic disorders in which specific / paroxysmal energy failure can impair function of any affected organ. Co-Enzyme Q10 and its analogues have shown benefit to patients with mitochondrial disorders. Mechanism of action is by serving as an electron carrier in the mitochondrial respiratory chain, as well as functioning as a potent lipid soluble antioxidant. It’s role is in minimising the effects of the disease on the child by supporting mitochondrial energy metabolism.</td></tr>
<tr><td>2. Details of medicine and indication
Please state whether licensed or unlicensed (off-label) use. Note that shared care is generally unsuitable for off-label prescribing unless it is a widely recognised use (e.g. included in BNF)</td><td colspan="2">Formulations: 30mg and 100mg capsules available from PharmaNord and other manufacturers.
Note that co-enzyme Q10 is not licensed for this indication and is licensed as a food supplement rather than a medicine.</td></tr>
<tr><td rowspan="3">3. Pharmaceutical aspects</td><td>Route of administration:</td><td>Oral, via nasograstric and PEG tubes</td></tr>
<tr><td>Formulation:</td><td>Capsules or Tablets available</td></tr>
<tr><td>Administration details:</td><td>Capsules to be pierced and capsule contents to be given orally, via nasogastric tube or PEG tubes. The capsule contents can be dissolved in water or fruit juice. Tablets can be crushed and mixed with a small amount of soft food such as yogurt/honey or jam.</td></tr>
<tr><td></td><td>Other important information:</td><td>• Do not prescribe as Lamberts or Nature’s Aid Co-Enzyme Q10 Capsules as they are blacklisted.
• Do not prescribe as liquid as it is a special which is prohibitively expensive.</td></tr>
<tr><td>4. Usual dose and frequency (including details of dose adjustments, e.g. in renal impairment) and duration of therapy
Transfer of monitoring and prescribing to Primary care is normally after the patient is on regular dose and with satisfactory investigation results. All dose or formulation adjustments will be the responsibility of the initiating specialist unless directions have been discussed and agreed with the primary care clinician.
The duration of treatment will be determined by the specialist, based on clinical response and tolerability.
Termination of treatment will be the responsibility of the specialist.</td><td colspan="2">The specialist will determine and advise on the dose to be used. 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.
Duration of treatment: Lifelong until no longer appropriate</td></tr>
<tr><td></td><td colspan="2">Baseline investigations</td></tr>
</table>

<table>
<tr><td>5. Baseline investigations and initial monitoring to be undertaken by specialist</td><td colspan="2">• 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.</td></tr>
<tr><td rowspan="2"></td><td>Monitoring</td><td>Frequency</td></tr>
<tr><td>• LFTs</td><td>• Will be taken and monitored by secondary care</td></tr>
<tr><td rowspan="2">6. Ongoing monitoring requirements to be undertaken by primary care</td><td>Monitoring</td><td>Frequency</td></tr>
<tr><td>• None</td><td>• Click or tap here to enter text.</td></tr>
<tr><td>7. Action(s) to be taken by primary care if abnormal result(s)</td><td colspan="2">• N/A</td></tr>
<tr><td>8. Cautions and contraindications
Please note this does not replace the Summary of Product Characteristics (SPC) and should be read in conjunction with it.</td><td colspan="2">Cautions
• May reduce insulin requirement in diabetes mellitus
• Reduce dose in moderate and severe hepatic impairment

Contraindications: N/A</td></tr>
<tr><td>9. Significant medicine and food interactions and management
For a comprehensive list, consult the BNF or Summary of Product Characteristics (SPC)</td><td colspan="2">• Ubidecarenone may reduce or enhance the effect of warfarin.</td></tr>
<tr><td rowspan="2">10. Adverse effects and management
Include details of incidence, identification, importance and management.</td><td>Adverse Effect</td><td>Action to be taken if detected</td></tr>
<tr><td>• Common or very common: diarrhoea, heartburn, nausea
• Rare: agitation/alertness/wakefulness, dizziness, headache, irritability</td><td>• Refer back to specialist if the patient has any side-effects
• 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.</td></tr>
<tr><td>11. Advice to patients and carers
The specialist will counsel the patient with regard to the benefits and risks of treatment and will provide the patient with any relevant information and advice, including patient information leaflets on individual medicines.</td><td colspan="2">• Advice will be provided by hospital initiating treatment.
• Further reputable patient friendly information can be found at: Ubidecarenone for mitochondrial disease – Medicines For Children</td></tr>
</table>

12. Pregnancy and breast feeding It is the responsibility of the specialist to provide advice on the need for contraception to male and female patients on initiation and at each review but the ongoing responsibility for providing this advice rests with both the GP and the specialist.	• Limited data but little to suggest this is a significant issue. Suggest discussion with metabolic team if planning for pregnancy of pregnancy arises unexpectedly. No specific contraception requirements.
13. Specialist contact information	

Dr. Toby Hunt	Tobias.hunt@nhs.net	RUH paediatrics
Metabolic Specialist nurses	ubh-tr.metaboliccns@nhs.net	BCH metabolic team
Dr Nicol West	Nicol.west@gwh.nhs.net	GWH paediatrics
Dr Jim Baird	jimbaird@nhs.net	SFT paediatrics 01722 336262 x 2584.
Other Specialist Contact Information		
• Click or tap here to enter text.		

14. Additional information For example, process for when Specialist or GP changes roles; specific issues related to patient age/ capacity/ specific monitoring.	• No specific monitoring requirements, local paediatrics teams/ BCH metabolic team will take responsibility for dose adjustments and continuation advice.
15. References	• Saudebray, 2011, Inborn Metabolic Diseases: Diagnosis and Treatment, 5th Edition, Springer. • British National Formulary for Children. Accessed online on 21/01/2022. • Guys and St Thomas', Kings college and University Lewisham Hospitals, Paediatric Formulary, 9th Edition, 2012 • Zschocke and Hoffmann, 2004, Vademecum metabolicum: Manual of metabolic paediatrics, 3rd Edition, Milupa Schattauer • Parikh et al, 2009, A Modern Approach to the Treatment of Mitochondrial Disease, Current Treatment • British Inherited Metabolic Disease Group. National Formulary for Inherited Metabolic Diseases. 2nd Edition 2020 Accessed online via BIMDG :: British Inherited Metabolic Disease Group on 21/02/22
16. To be read in conjunction with the following documents	• NHS England: Responsibility for Prescribing Between Primary & Secondary/ Tertiary Care. Ref 07573, Version 1.0, Published January 2018. Accessed via: https://www.england.nhs.uk/publication/responsibility-for-prescribing-between-primary-and-secondary-tertiary-care/

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