1. Introduction and Who Guideline applies to

1.1. These Clinical Guidelines have been developed in order to assist and ensure consensus is maintained among Senior Specialist Paediatric Ketogenic Dietitians and Senior Paediatric Ketogenic Dietitians, implementing ketogenic diet (KD) treatment for paediatric patients with intractable epilepsy.

This Clinical Guideline is intended to be used for the management of children aged 0-16 years if accessing standard education or those aged 0-19 years if requiring special needs education. It will also act as a source of reference for the Paediatric Ketogenic Diet Multi-Disciplinary Team (MDT). **A patient cannot undertake a KD unless they are under the care of the Paediatric Ketogenic MDT.** This Clinical Guideline is not to be used by Dietitians or other Health Care Professionals to start a patient on KD in isolation from the Paediatric Ketogenic Diet MDT.

The Paediatric Ketogenic Diet MDT

- Consultant Paediatric Neurologist
- Senior Specialist Paediatric Dietitians (Band 7)
- Senior Paediatric Dietitian(s) (Band 6) on KD rotation
- Paediatric Neurology Specialist Nurse

The KD is a high fat, adequate protein and very low carbohydrate diet that mimics the metabolic state of starvation. In the absence of adequate glucose supply, ketone bodies; acetoacetate and β-hydroxybutyrate become the main energy source for the brain. The exact mechanism of action of the KD is not fully known, but there are a number of hypotheses; KD increases the energy reserves of the brain which may increase the production of inhibitory neurotransmitters; there is increased glutamate available for the synthesis of gamma amino butyric acid (GABA) which is the main inhibitory brain neurotransmitter; KD may have anti-inflammatory properties that in turn suppresses seizure activity [1]; medium-chain fatty acids, a common component of ketogenic diets, have been shown to directly inhibit AMPA receptors potentially blocking seizure onset and raising seizure threshold [2]; the acetoacetate ketone bodies may have a direct anticonvulsant effect, and the kilocalorie (kcal) control may also contribute to the anticonvulsant effect of the KD.

The KD was first used as a treatment for epilepsy in 1921, after it had been incidentally noted that fasting patients with epilepsy had fewer seizures [3]. Many studies have reported effectiveness [4, 5, 6, 7]. One randomized controlled trial [8] reported a 75% reduction in seizure activity in the group of children treated with KD after 3 months on diet. 28 children (38%) experienced greater than 50% reduction in seizure activity and 5 children (7%) experienced greater than 90% seizure reduction. Initially, the KD is used in addition to the patient's usual anti-epileptic drugs (AED's), however it is possible that the AED's may be reduced or stopped if the diet is successful.

Aim of Dietary Treatment

- To calculate a paediatric KD prescription that will provide optimal seizure control and maintain adequate nutrition for growth.

Objectives

- Reduce frequency of epileptic seizures
- Reduce severity of epileptic seizures
- Meet the patient's nutritional requirements
- Ensure adequate nutrition for growth
- Minimize the occurrence of complications e.g. poor tolerance of the KD
- Ensure cost effective use of prescribable nutrition & diet products

1.2. This Clinical Guideline is intended for use by Senior Specialist Paediatric Dietitians (Band 7) and Senior Paediatric Dietitian(s) (Band 6) on the ketogenic diet rotation working within the Paediatric Ketogenic Diet MDT.

1.3 The aim of this Clinical Guideline is to ensure clarity and consistency on the use of KD for paediatric patients. It sets out the appropriate procedures to be followed;

- When choosing the type of KD
- Calculating the individual KD prescription
- Initiating KD
- Fine tuning KD to optimise ketosis
- Monitoring patients while on KD
- Weaning patients from KD

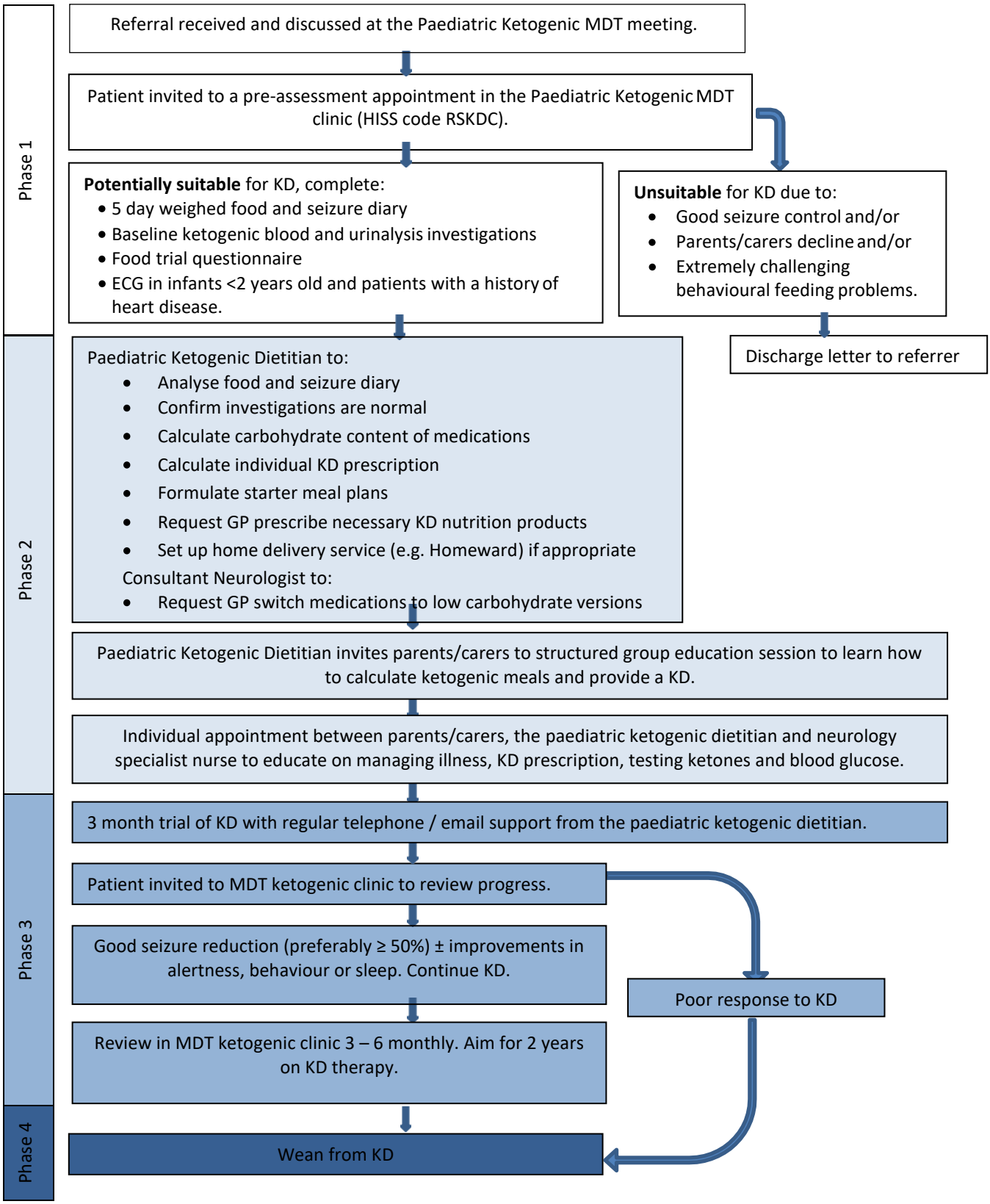
1.4 This Clinical Guideline will be used predominantly for the treatment of paediatric outpatients. However, it will also guide on the procedures to follow when a paediatric patient is admitted to hospital.

Indications for a planned admission include:

- Any infant under 1 year old starting KD
- Children starting KD at greater risk of hypoglycaemia or hyperketosis e.g. history of idiopathic hypoglycaemia.

2. Guideline Standards and Procedures

2.1. Paediatric Ketogenic Diet care pathway



2.2. Phase 1 – Referral and Assessment

a) Referral Criteria

The Consultant Paediatric Neurologist refers the patient to the Paediatric Ketogenic MDT Service if; they have intractable epilepsy [9, 10,11], have a Glucose Transporter Type 1 (GLUT-1) deficiency or Pyruvate Dehydrogenase deficiency (PDH).

- Contraindications to trialling KD include the following [9]:

- Absolute Carnitine deficiency (primary)
  - Carnitine palmitoyltransferase (CPT) I or II deficiency
  - Carnitine translocase deficiency
  - b-oxidation defects
  - Medium-chain acyl dehydrogenase deficiency (MCADD)
  - Long-chain acyl dehydrogenase deficiency (LCADD)
  - Short-chain acyl dehydrogenase deficiency (SCADD)
  - Long-chain 3-hydroxyacyl-CoA deficiency
  - Medium-chain 3-hydroxyacyl-CoA deficiency
  - Pyruvate carboxylase deficiency
  - Porphyria

Relative

- Inability to maintain adequate nutrition
- Surgical focus identified by neuroimaging and video-EEG monitoring
- Parent or caregiver noncompliance
- Propofol concurrent use (risk of propofol infusion syndrome may be higher)

Referral letters should include the following information;

- The patient's present medical condition and medical history
- Seizure type, severity and frequency
- Previous medication tried
- Dosage of current medication
- Patients feeding ability and route for nutrition (oral or enteral tube)
- Anthropometry including weight and a summary of any growth concerns.
- Information on recent investigations e.g. Electroencephalogram (EEG)
- If the patient recently had or is awaiting Vagus Nerve Stimulation (VNS)
- If the patient is a candidate for neurosurgery
- And meet basic standards of documentation as outlined by the 'Policy for Documenting in Patients' Health Records (in all media)' Trust Ref B30/2006.

b) Ketogenic Pre-assessment clinic

New patient referrals can be discussed in the monthly paediatric KD MDT meeting. The patient and parents/carers are invited to attend a forty five minute clinic pre-assessment appointment with the paediatric KD MDT. The following information is collected and considered by the Consultant Paediatric Neurologist:

- Epilepsy syndrome and associated symptoms

- Other diagnosis
- Anticonvulsants tried and current medications
- Current seizures – type, frequency, severity and semiology of each seizure type
- Presence of any behavioural issues
- Previous EEG and Magnetic resonance imaging (MRI) findings
- Information for parents on mode of action of KD, seizure reduction, medication reduction, cognition improvement, possible side effects and the length of treatment

The **Senior Specialist/ Senior Paediatric Ketogenic Dietitian** records and plots the patient's current weight and height and assesses if poor weight gain has been a concern in the past. If the patient has a nasogastric tube or gastrostomy tube insitu it is useful to liaise with the patients home enteral feeding dietitian for information regarding past growth, tolerance to feed, feeding plan and goals.

The **Senior Specialist/ Senior Paediatric Ketogenic Dietitian** will take a brief diet history to assess current dietary intake, in particular to establish if the patient is very fussy with food. The dietitian will also advise the parents/carers regarding:

- What a ketogenic diet is
- The prescriptive nature of the diet
- The importance of accurately weighing food
- The need for regular weighed food and seizure diaries
- External support groups/resources available e.g. Matthews Friends charity
- Information starter pack provided

The **Neurology Specialist Nurse advises** the parents regarding:

- Monitoring of ketones and blood sugars
- Brief description on use of the blood ketone and glucose meter
- Individual seizure care plans for administration of prescribed rescue medication e.g. Buccolam.

The **Consultant Paediatric Neurologist** advises the parents regarding:

- The indications for and aims of KD therapy
- The need for regular bloods and urinalysis
- Potential side effects of the KD and associated risks
- Duration and discontinuation of the diet
- Ketogenic Pathway - what's next if the family decides to pursue KD?

The **KD MDT and parents/ carers** agree together if KD is feasible or not for the patient. If KD is not a viable treatment option the child is discharged back to their referring Consultant Paediatric Neurologist. If the decision is to pursue KD, a starter information pack will be provided. Parents should book a bloods appointment for their child with the Children's Outpatient Department, Leicester Royal Infirmary (LRI). The Senior Specialist Paediatric Ketogenic Dietitian will complete the blood and urine order forms and give these to the parents/carers to be taken to this appointment. See Appendix 1.

The following Written information/Resources are provided in the starter pack:

- Contact details for the KD Team
- 'Next Steps Before Undertaking a ketogenic diet'
- 'A guide to the modified ketogenic diet' OR 'The classical Ketogenic Diet' depending on the type of KD the child is most likely to undertake
- 'The initial Ketogenic Foods Trial' questionnaire if orally fed
- 'I am going on Ketogenic Diet' – produced by Matthews friends charity
- 'Introduction to Medical Ketogenic Therapies for Children' – produced by Matthews Friends charity.
- A weighed food and seizure diary to be completed and returned within an agreed deadline, 3 weeks for example
- A urine pad (if appropriate) and urine bottle
- Consent to contact by email form

Parents/ carers are advised to visit the Matthews Friends website www.matthewsfriends.org. This is a charity organisation that supports parents/ carers and patients on the various ketogenic diets. They provide recipe ideas, cookery demonstrations, and practical advice regarding living with ketogenic diets, parent's forums, useful literature, education days, and general support for families on ketogenic diets, all free of charge.

c) Follow up telephone call

The Senior Specialist / Senior Paediatric Ketogenic Dietitian telephones/ emails the family in the weeks following pre-assessment clinic (if no contact has already been made by the family) to answer any further questions and follow up on progress with the weighed food and seizure diary etc.

2.3. Phase 2 – Preparation

Use the ketogenic diet new starter checklist (see appendix 2) to ensure all necessary tasks are completed before KD starts.

a) Essential tasks before KD can commence

Tasks to be completed by parents:

- Return a 5 day weighed food and seizure diary
- Anthropometrics – Recent clinic weight & length/ height can be used if appropriate, if not the patient should be measured locally if possible; at school or GP surgery. If not it can be arranged at the LRI.
- Pre diet electrocardiogram (ECG) in infants <2 years old and patients with a history of heart disease [9]
- Blood investigations, listed in Appendix 1
- Urinalysis investigations, listed in Appendix 1
- Attendance at a structured group education session on KD meal calculation using 'choices' for oral diet. Calculation of Classical KD recipes can be demonstrated using an electronic ketogenic calculator e.g. electronic ketogenic manager (EKM) programme.
- Completion of a worksheet calculating basic meals with the KD foods choices lists or simple meals in EKM

- Parents/ Carers are confident in meal calculation and agree to undertake calculation of meals for their child
- Parents/carers to complete an education session with the paediatric neurology specialist nurse to become competent at monitoring serum ketones and blood sugars at home
- Where changes to AED medication is needed (i.e. changing from liquid to tablet formulation) this should be commenced at least 1 week prior to starting KD to ensure no adverse effects on seizure control from this change

Tasks to be completed by Senior Specialist/ Senior Paediatric Ketogenic Dietitian to enable KDto commence:

- Food and seizure diary to be analysed using nutritional analysis software e.g. Nutritics
- Calculation of carbohydrate content of medications and discuss necessary changes with Consultant Paediatric Neurologist and Parents/ Carers
- Confirm appropriate KD and calculation of KD prescription
- Calculate appropriate dose of FruitiVits
- Arrange a group education session for the family
- Devise suitable KD meal plans
- Request starter stock of prescribed nutrition products e.g. Liquigen or FruitiVits from manufacturer sample services if parental consent has been obtained to share necessary details
- Letter to GP to request KD prescription nutrition products
- Arrange for delivery of feed products for children who will receive ketogenic diet via a feeding tube
- Check bloods and urinalysis results and report any abnormalities to Consultant Paediatric Neurologist
- Confirm that the Consultant Paediatric Neurologist is happy for patient to commence KD

Tasks to be completed by Consultant Paediatric Neurologist to enable KD to commence:

- Write to the GP to confirm that the patient is ready to start KD and request the necessary changes to medications to reduce carbohydrate intake
- Where the patient is under 6 years old, write to the parents to advise them on the appropriate doses of the low carbohydrate paracetamol formulation Calpol Six Plus 250mg Fastmelts and low carbohydrate ibuprofen formulation Nurofen 200mg Meltlets.

b) Food and SeizureDiary Analysis

A food & seizure diary is provided at the pre-assessment clinic. All food, drinks, medications and oral nutritional supplements taken should be recorded along with all seizure activity for 5 days. This should be returned to the Senior Specialist / Senior Paediatric Ketogenic Dietitian within 1 month. If it is not returned within that time the Senior Specialist / Senior Paediatric Ketogenic Dietitian writes to the parents/ carers to check if they wish to continue, if so the food diary must be returned within a further 2 weeks from that date. The parents/ carers are reminded that the patient will be discharged from the ketogenic diet service and removed from the waiting list, if a food and seizure diary is not received.

These time limits can be extended at the discretion of the ketogenic diet MDT/ Dietitian. The accuracy of the food and seizure diary is a good indication of the parental commitment to undertaking the KD. If the food and seizure diary is incomplete, the parents/ carers may be requested to complete a further food and seizure diary, with clear advice on how to improve this from the Senior Specialist / Senior Paediatric Ketogenic Dietitian. The Senior Specialist / Senior Paediatric Ketogenic Dietitian analyses the food diary to assess usual energy intakes (using nutritional analysis computer software).

c) Calculation of carbohydrate content of medications

The carbohydrate content of all of the patient's regular medications should be checked. The local Medicines Information Service can provide the carbohydrate level for medications; however this can take weeks in some instances. They will require the name, brand, manufacturer, dose, preparation (liquid/tablet) and concentration if applicable of the medication in question. Any medications containing carbohydrate should be discussed with the Consultant Paediatric Neurologist and if possible changed to a crushable/dissolvable tablet preparation to reduce the carbohydrate content. The Consultant Paediatric Neurologist will inform the General Practitioner (GP) in writing of any required medication changes and request that these prescriptions continue as appropriate. If the medication must continue, the carbohydrate it contains should be included in the patient's daily carbohydrate allowance, particularly if the total carbohydrate from medications exceeds 1 gram per day.

d) Choosing and calculating KD

The Senior Specialist Paediatric Ketogenic Dietitian in discussion with the parents/ carers chooses the most suitable KD for the patient; the different types of KD are described in Appendix 3. The KD energy prescription will be derived from an average of the energy intake from the food diary and the child's estimated average requirement (EAR) for energy.

Appendix 4 details the products used with KD. An individual dietary prescription is calculated for the patient, taking account of the information obtained from the food diary analysis. Appendix 5, and 6 provide examples of calculating the dietary prescriptions and meal plans for the Classical KD and the modified KD respectively.

e) Calculation of Fruitivits dose

Most children require between half to a full sachet of FruitiVits daily. Calculate the dose that meets the Recommended Nutrient Intake (RNI) for the main vitamins and minerals such as calcium, iron, selenium, zinc, copper and B vitamins. Remember to take account of the vitamin A content of double cream particularly if calculations include this as a dominant fat source and the calcium contribution from the child's milk allowance. FruitiVits contain negligible potassium, sodium and chloride so enteral feed regimes (particularly for those patients with very low kcal requirements or on a modular feed recipe) may require additional electrolyte supplementation. Discuss this with the Consultant Paediatric Neurologist. Always aim to meet at least the Lower Reference Nutrient Intake (LRNI) for vitamins and minerals.

f) Structured education group session – Modified KD – Food choices & calculation of meals

Those families who decide to pursue KD will be invited to an education session (ideally 3-4 sets of parents/ carers per session). Where possible the individual KD diet prescriptions should already be calculated for each child so that the parents/ carers can work on meal plans applicable

to their child rather than a hypothetical KD diet prescription. Ideally both parents/ carers and/ or somebody who will be supporting either parent/ carer should be encouraged to attend. This support system will be essential for the parent/ carer if their child moves onto KD. The patient does not usually attend but can if appropriate (i.e will not provide a distraction and can engage with the education session).

This session will cover the pre-defined food lists and calculation of meals in more depth. Parents/ carers need to be able to demonstrate that they can use the food lists for simple meals, otherwise their child may not be able to undertake KD. Individual cases where parents/carers struggle may be accommodated, where the Senior Specialist / Senior Paediatric Ketogenic Dietitian needs to undertake all meal calculations for the patient for the duration of the KD. However, the Senior Specialist / Senior Paediatric Ketogenic Dietitian cannot commit to doing this for all patients so it is essential it is reserved for **exceptional circumstances only.**

Worksheets will be given to the parents to be completed and returned to the Senior Specialist / Senior Paediatric Ketogenic Dietitian to demonstrate parent/ carer ability to undertake KD. These will involve the calculation of basic meals. Parents/ carers will be encouraged to introduce higher fat foods to their child's diet and remove refined carbohydrates like crisps, sweets, chocolate, fizzy drinks and biscuits. Parents will be encouraged to trial preparing some ketogenic meals at home, weighing all ingredients. It is important that the parents/ carers expectations are discussed; the Senior Specialist / Senior Paediatric Ketogenic Dietitian should facilitate this discussion and encourage realistic expectations.

Parents/ carers are asked to complete a confidence questionnaire, assessing their confidence to undertake KD before the education session and then after the session.

The following written information will be provided in a pack:

- The Modified KD food choices lists (laminated)
- Starter plan including illness management
- A copy of the PowerPoint presentation slides
- Label reading calculation sheets
- Calculating Ketogenic meals worksheet
- Daisy's Keto Café postcard (Daisy Garland charity)
- Quick User Guide to ordering online (Nutricia Homeward) (if appropriate)
- Non prescribable product list (Ketogenic Dietitian's Research Network (KDRN))
- Sweetener information (KDRN)
- "Me and my Ketogenic Diet. Luna's story" (Nutricia)
- Admission information cards

g) Individual/structured education group session for those undertaking classical KD including the use of EKM (electronic ketogenic calculator)

Classical KD is mainly used in those children who are solely enterally tube fed or for those who have a mixed intake – some oral intake and some enteral intake via a nasogastric tube or gastrostomy. The classical KD is also recommended for children under 2 years old who are starting KD [10]. The classical KD is recommended in 0-2 year olds because of the difficulty in tolerating high doses of MCT fats and the need for strict control. If the patient is to commence the Classical KD and takes foods orally; request a copy of EKM from the developer (check Matthews's Friends website, http://www.matthewsfriends.org for the most up to date version and

contact details for the developer) for the family.

Access to this programme will only be granted with permission from a Senior Specialist / Senior Paediatric Ketogenic Dietitian. EKM is free to Dietitians and families. The Senior Specialist / Senior Paediatric Ketogenic Dietitian should provide an EKM user's manual (supplied by Matthews Friends) and undertake an education session with the parents demonstrating the use of EKM. 'You tube' videos are available to support this training.

The following written information will be provided in a pack;

- A guide to EKM
- The classical KD food choices lists
- A copy of the PowerPoint presentation slides
- Starter plan including illness management
- A guide to prescribed products (if appropriate)
- Label reading calculation sheets
- Daisy's Keto Café postcard (Daisy Garland charity)
- Quick User Guide to ordering online (Nutricia Homeward) (if appropriate)
- Non prescribable product list (Ketogenic Dietitian's Research Network (KDRN))
- Sweetener information (KDRN)
- "Me and my Ketogenic Diet. Luna's story" (Nutricia)
- Admission information cards

h) Calculating ketogenic meals

The Senior Specialist / Senior Paediatric Ketogenic Dietitian aims to calculate ketogenic versions of the child's favourite meals incorporating typically low carbohydrate fruit and vegetables where possible. Ideally protein, fat and carbohydrate should be spread evenly throughout the day. Where possible there will be identical choices at lunch and dinner so parents can batch cook and use these meals interchangeably. The MCT fat source, should be divided into regular daily doses and guidance given on how to incorporate it into the diet. It is best to continue with the child's typical meal plan, for example if s/he usually has 3 meals and 1 snack then continue with this.

Sufficient options to provide a 3-5 day meal plan will be prepared to help start the patient on KD. This gives parents/carers examples of appropriate meals, which they can alter as they become more confident.

i) MCT fat introduction

MCT fat can increase ketone levels efficiently and should be trialled in all KD regimens. Where high volumes of MCT fat is used (e.g. more than 25g MCT per day in children over 5 years old) the MCT source should be introduced slowly over approximately 3-5 days due to potential for MCT fats to cause gastrointestinal disturbance. This is usually done with Liquigen rather than MCT oil as it is more versatile. For the MKD and oral classical KD this is usually done by gradually introducing one KD meal or snack with liquigen per day. During this time the parents can try incorporating it into their child's diet in a number of different ways to ascertain which ways they will take it, for example in milk, no added sugar squash, food, from a syringe or in sugar free jelly. Parents should be encouraged to start preparing for the full KD to commence, e.g. shopping and meal planning.

2.4. Provision of specialist products for KD

a) A starter supply of nutritional products can be requested from manufacturer sample services or Daisy Garland KetoCafe welcome boxes by the Senior Specialist / Senior Paediatric Ketogenic Dietitian if parental consent has been obtained to share the necessary details. For example, request 3 x 250ml bottles of Liquigen and sachets of FruitiVits.

If the patient is enterally tube fed they should already be registered with Nutricia Homeward. The patient must be registered to receive delivery of Ketocal (enteral feed). Ketocal cannot be dispensed through a pharmacy; it must be delivered via Nutricia

Homeward. Polycal, Protifar, Fruitivits, Complete amino acid mix, Optifibre, Calogen, MCT Oil and/or Liquigen can also be ordered through Nutricia Homeward. The Senior Specialist / Senior Paediatric Ketogenic Dietitian requests in writing that the patient's GP prescribe all of the necessary nutrition and dietetic products. The GP should provide a copy of that prescription to Nutricia Homeward for each order and delivery. This process can be significantly delayed so do not commence KD using a sample supply of products, wait until the patient has received a home delivery of all the products necessary to undertake KD.

The paediatric neurology specialist nurse keeps a supply of blood glucose and ketone testing meters and ketone test strips. The local area representative (e.g. Abbot FreeStyle Optium Neo) will provide further supplies of the meter when depleted free of charge.

Each patient should be supplied with:

- Blood ketone and glucose meter which usually includes 10 lancets and 10 blood glucose test strips
- Box of 1 x 10 ketone test strips specific to the meter supplied.

The paediatric neurology specialist nurse should educate the parents/carers on the use of this equipment before starting KD. The paediatric neurology specialist nurse should request in writing that the child's GP make ongoing prescriptions of the test strips and lancets specifying the quantities required. In addition the GP should provide a sharps box.

b) Starting KD plan for parents

This is a time line for the parents. It advises parents on how to gradually introduce ketogenic meals. It will also advise on when to commence ketone and blood glucose testing and when to commence FruitiVits.

c) Blood and urinalysis results

The Senior Specialist / Senior Paediatric Ketogenic Dietitian checks the baseline KD blood and urinalysis investigations once complete, reporting any concerns or abnormalities to the Consultant Paediatric Neurologist. Once all are reported the Consultant Paediatric Neurologist checks the investigations and confirms that the child can commence KD. The Senior Specialist/ Senior Paediatric Ketogenic Dietitian then communicate that decision to the family.

2.5. Phase 3 – Initiating and maintaining ketogenic diet

a) Commencing KD

A three month trial of KD is undertaken initially to assess its effect on seizure control. It is standard practice in the UK and here at University Hospitals of Leicester NHS Trust (UHL) to initiate KD in the outpatient setting. Exceptions include starting KD in children under 1 year old for whom it is recommended to start KD in the hospital setting [10]. Where there are no complications this admission would be expected to be 1-2 weeks long. If necessary, older children can be admitted, particularly if the family require intensive support and education or there are concerns over risks of hypoglycaemia. Appendix 7 provides further information on managing inpatient KD initiation. If the patient is to start as an outpatient, the parents/ carers are invited to another outpatient appointment with the Senior Specialist / Senior Paediatric Ketogenic Dietitian, during which the following will be discussed/ demonstrated and written information provided:

- Getting started on KD – time line for introduction of KD including Liquigen
- When to check serum ketones and blood glucose and acting on results
- Managing illness whilst on KD and recognising hyperketosis and hypoglycaemia
- Weekly seizure record sheet
- The KD prescription and meal plans
- The FruitiVits dose
- Recipe ideas
- A copy of the letter sent to the GP requesting prescribed KD products
- A letter from the Consultant Paediatric Neurologist advising on appropriate doses of over the counter low carbohydrate paracetamol and ibuprofen for children under 6 years old.
- Parent/ carer asked to give signed consent for use of Nutricia Homeward delivery service, as required

The paediatric neurology specialist nurse demonstrates blood ketone and blood glucose testing to the parents/ carers and provides written information to support this. Blood ketone testing is used in preference to urine ketone testing because urine testing is less accurate and cannot indicate hyperketosis (>16mmol/l). Blood glucose should be tested for at least the first two weeks while initiating KD and if normal then monitoring can stop. Appendix 8 details the daily monitoring that parents/ carers undertake and appendix 9 describes the treatment of hypoglycaemia and hyperketosis.

KD should be started at the weekend by introducing one meal or snack per day until the child is on full KD. By starting on a weekend the child should reach full KD on a week day when dietetic support is available. No changes should be made on a Friday – Sunday because dietetic support is not available at the weekend. The child undertakes a 3 month trial of KD. If there is no improvement in seizure control or the diet is not tolerated, the patient is then weaned off the KD (see section 3.8 and appendix 10).

All relevant agencies involved with the patient should be informed that s/he has commenced KD, e.g. consultant paediatric neurologist and GP. It is primarily the responsibility of the parents/ carers to ensure that school/nursery, respite centre and any informal childcare providers are informed.

2.6. Hazard alerts identification list

Every patient on KD must have a 'Hazard Alert Identification List' form filed in the front of their medical notes with a red alert sticker on the front cover. The Senior Specialist / Senior Paediatric Ketogenic Dietitian is responsible for ensuring this task is completed. Search for this form by putting the title into UHL intranet search engine, to ensure that the most up to date version is being used. Attach a copy of 'Management of Children on Ketogenic Diet for Epilepsy' Trust Ref: C255/2016 to the hazard alert form.

In the 'Allergy/Sensitivity' 'other' section, record –

"On Ketogenic diet for epilepsy. - If admitted to LRI, contact the paediatric neurology team via switchboard and the Senior Specialist Paediatric Ketogenic Dietitian (ext 15400)

- *Maintain blood glucose >2.5mmol/l and ketones 2.0-5.0mmol/l.*
- *If IV fluids are required use normal saline if the blood glucose is >2.5mmol/l and ketones are 2.0-5.0mmol/l.*
- *Use 5% dextrose with 0.9% saline solution if blood glucose is <2.5mmol/l and/or ketones are >5.0mmol/l"*

This 'Hazard Alert Identification List' form should be crossed through with the date and signed when the patient is no longer on KD.

2.7. Monitoring patients while on ketogenic diet

The Senior Specialist / Senior Paediatric Ketogenic Dietitian contacts the parents/ carers regularly via telephone or email. This can involve daily contact when starting KD decreasing to weekly then fortnightly reviews as required when established on KD. Parents/carers are encouraged to contact the Senior Specialist / Senior Paediatric Ketogenic Dietitian as needed via email or telephone. Frequent contact is essential to ensure the family feels supported and problems are dealt with swiftly. Compliance, tolerance and ketosis are monitored during these contacts. Fine tuning of the diet prescription is undertaken in response to poor ketosis or poor tolerance.

The Senior Specialist / Senior Paediatric Ketogenic Dietitian has limited access to a clinic room in the children's outpatients department however families can be seen at hospital or at home if they are having difficulties beyond what can be dealt with via telephone/video call. The patient is requested to attend a clinic with the KD MDT at 3 months and 6 months on KD and then every 6 months while they continue on KD. Children under 2 years of age may require an additional appointment at 6 weeks after starting KD, and should attend KD MDT clinic every 3 months until the age of 2 years, after which they can be seen every 6 months [10].

At each clinic appointment the following are reviewed;

- Seizure control and level of ketosis
- Medications
- Compliance with KD
- Anthropometrics
- Side effects and tolerance of KD
- Blood and urinalysis results including nutritional bloods
- Need for further investigations e.g. renal ultrasound, EEG
- Social; effect on schooling/ respite or going on holiday

a) Seizure Control

The parent/ carer is asked to complete a 5 day food/ feed and seizure diary at pre-assessment, 3 months on KD, 6 months on KD and every 6 months thereafter (or every 3 months for children under 2 years). This can help to give some understanding of seizures when seizure activity is daily and visually apparent. The patient's seizure records are analysed for any changes in seizure activity. It is hoped that the patient will experience some degree of seizure reduction. Other benefits reported by parents E.g. increased alertness, improved sleep pattern, improved behaviour will be taken into account when considering if KD has been successful. If ketone levels are lower than ideal (2- 5mmol/L) and seizure control is not considered to be optimal then fine tuning of the diet will be undertaken as discussed below.

b) Medications

The patient's anti- epileptic drugs (AED's) are checked including doses and timings. Side effects to these AED's are monitored. If the KD has had a positive effect and significantly reduced seizure activity, the Consultant Paediatric Neurologist may consider reducing the medications.

c) Compliance

The KD is a complex diet to follow which can prove difficult for many families. It impacts greatly on the family which in turn can often influence compliance with the diet. The Senior Specialist / Senior Paediatric Ketogenic Dietitian explore these issues with the family and offers practical ideas to overcome these problems in the future. The Senior Specialist / Senior Paediatric Ketogenic Dietitian checks if the patient appears satisfied with their meal plans and/ or complains of hunger. If so, an increase in the energy prescription may be indicated, particularly if growth appears to be inadequate. If growth is adequate and an energy increase is not indicated; changing the meal plan to include smaller, more regular meals and snacks may help with hunger.

d) Anthropometrics

The patient's weight and length/ height are measured when they attend clinic. It is important to remember that patients on the classical KD are not eating to hunger; they are eating to a set energy prescription based on an average of their food diary analysis and estimated average energy requirement. We monitor weight gain very closely as it is well documented that weight gain reduces while on KD [12, 13, 14], although in practice this is not a regular observation. Height is initially maintained while on KD but the longer a patient stays on KD, particularly younger children, the greater the risk of them not following their height percentile [12]. It has been demonstrated that catch up growth (height) to baseline percentile occurs, once weaned from KD [14].

e) Side effects and tolerance

The potential side effects of KD are often grouped into shorter and longer term side effects [15, 16, 17]. The short term side effects include lethargy, nausea, vomiting, diarrhoea, constipation and abdominal cramping. These can be managed in a variety of ways to improve the patient's tolerance:

Slow initial introduction of KD and in particular MCT fat. Reduction in MCT fat may be necessary

for a short period of time to improve tolerance. Once symptoms resolve, the MCT fat can be increased again but more slowly. Always give the MCT fat source (Liquigen/ MCT oil) with a meal or snack to aid tolerance. Try using different fat sources; some children have struggled with the level of double cream and butter on the Classical KD but tolerated Calogen well as a substitute.

Constipation on KD is common due to the low fibre intake. Adequate fluid intake should be encouraged. Where symptoms of abdominal cramping, nausea, vomiting and/or diarrhoea are reported constipation ± overflow diarrhoea should be considered. A low carbohydrate laxative such as Movicol should be considered.

Longer term side effects include the risk of slow growth described previously, dyslipidaemia [18] and the development of renal stones [19], both discussed below.

f) Blood and urinalysis results

See Appendix 1 for a list of routine blood and urinalysis tests. Results are reviewed and discussed with the parents/ carers. Incomplete blood and urinalysis investigations may need to be repeated, particularly if a parameter is missing that was previously abnormal.

The results can indicate a need for modifications to the KD prescription or vitamin and mineral dose (FruitiVits, electrolytes). The Senior Specialist / Senior Paediatric Ketogenic Dietitian will aim to manage these modifications where possible through the use of food and dietary changes; however additional nutritional supplements or an increase in the vitamin and mineral preparation may be necessary. For example, if the lipid profile is worsening, the Senior Specialist/ Senior Paediatric Ketogenic Dietitian will advise parents to choose 'healthier fats', replacing saturated fats like butter and double cream with monounsaturated fats like olive oil. Introduction of rich food sources of particular micro nutrients may be indicated, for example very small doses of Brazil nuts (one every few days or 3 per week depending on age of child and blood result) will help to increase intake of Selenium and Potassium.

FruitiVits should be increased with caution. It should not be increased in response to one low blood parameter as increasing the dose to correct this low parameter may result in excessive levels of other micro-nutrients such as vitamin A. A detailed nutritional analysis should be undertaken using nutritional analysis software, comparing the results with known toxicity levels for various micro- nutrients, particularly the fat soluble vitamins. In addition, it is essential to ensure the patient is meeting at least their LRNI for all micronutrients but in particular electrolytes. If not then prescribed electrolyte supplements should be considered. Discuss this with the Consultant Paediatric Neurologist and together decide on a suitable preparation and dose. Alternatively where potassium and sodium intake is low, the Senior Specialist / Senior Paediatric Ketogenic Dietitian could advise on an appropriate daily dose of a potassium chloride based salt substitute e.g. LoSalt (Klinge Foods Ltd). Electrolyte intake from medications (e.g. sodium valproate, potassium citrate) must be taken into account when considering electrolyte supplementation.

Blood results can indicate anaemia and dietary modifications should be advised within the restrictions. However, iron supplementation is likely to be necessary. The Consultant Paediatric Neurologist arranges prescription of this.

Vitamin D levels can be insufficient both before KD and on KD and supplementation will be recommended. The Consultant Paediatric Neurologist arranges prescription of these products.

Many patients on KD experience a raised urinary calcium:creatinine ratio, indicating an increased risk of renal stones. The calcium:creatinine ratio can be reduced back into the normal range by treating with Potassium Citrate. The Consultant Paediatric Neurologist will arrange prescription of this.

A small number of patients have become Carnitine deficient. Carnitine is essential for the transport of fat to the cells to be broken down. MCT fat does not require a Carnitine shuttle, long chain triglyceride (LCT) fat only is transported in this way. The Consultant Paediatric Neurologist will arrange prescription of Carnitine if required.

g) Further investigations

It is estimated that there is a 5-6% risk of developing renal stones while on KD, due to chronic acidosis, urine acidification, hypercalciuria and hypocitraturia [19]. A raised calcium creatinine ratio indicates hypercalciuria. Patients with a raised calcium creatinine ratio will usually undergo a renal ultrasound to check for evidence of renal stones. The Consultant Paediatric Neurologist will decide if this is necessary and arrange it. As a minimum each patient will have an annual renal ultrasound scan.

h) Social

The patients school and/or respite centre often require support from the KD team to enable them to confidently care for the patient. Parents/carers often request that we liaise with the school/respite centre for various reasons. The patient's progress in school is discussed and in particular if there are any reported improvements in behaviour, alertness or cognition. Families can be reluctant to undertake 'usual' activities like school trips or holidays, the Senior Specialist / Senior Paediatric Ketogenic Dietitian and Neurology Specialist Nurse can support them with preparing for these activities.

If flying abroad, a letter will be necessary for the airline to justify the need for additional hold baggage and fluids (such as Liquigen) to be taken into the cabin.

The paediatric ketogenic dietitians will intermittently run education sessions for schools/ nurseries/ respite centres of patients on the paediatric KD caseload and patients planning to start a ketogenic diet.

2.8. Fine Tuning the KD

Fine tuning of the initial KD prescription is always expected, to achieve optimal ketosis. The most likely reason for poor ketosis is that the percentage fat or fat ratio is not high enough and requires adjustment. However illness, poor compliance with the diet prescription and over- or underfeeding can all affect ketosis.

a) If following the Modified KD – Consider if:

- Excessive amounts of protein is being eaten
- Carbohydrate content of high protein foods is accounted for e.g. beans, lentils, nuts
- Parents/carers are calculating the fat choices and carbohydrate intake correctly
- There are any new carbohydrate containing medications
- Appropriately low carbohydrate drinks are being used
- All fat choices are being consumed

- The correct volume of MCT fat is being given.
- The kcal prescription istoo high
- There are a lot of processed foods being used and miscalculated
- There are a lot of low carbohydrate alternative foods which are very high in protein
- Meals are being finished particularly the fat source
- Constipation is affecting appetite and compliance

When the above issues have been considered and addressed but ketones remain low the fat and/ or carbohydrate prescription needs adjusting. Usually the carbohydrate content on the modified KD will already be minimal and further reduction can negatively affect palatability and acceptance of thediet.

The fat intake could be increased to increase ketone levels but the change to total kcal intake should be considered. Excessive kcal intake can in turn have a negative effect on ketosis. An alternative option is to replace some LCT fat with MCT fat to improve ketosis without increasing kcal intake. MCT should be increased gradually.

Where the high fat meals are not well tolerated ketogenic oral nutritional supplements could be considered to reduce the fat content of meals.

It may be useful to request a weighed food diary to assess the protein intake to ensure that this is not excessive. High protein intake could reduce the KD ratio or cause reduced appetite for prescribed fat intake.

b) If following the Classical Ketogenic Diet – Consider if;

- Meals are being calculated correctly using the electronic calculator
- All foods are being weighed and not estimated
- Exact macronutrient quantity is being used and not rounded up e.g. 3.5g carbohydrate, not 5g.
- Meals are being finished particularly the fat source
- Enteral feeds are made up correctly according to the recipe
- Where there is an option to give oral or enteral tube feeds, appropriate amounts are given
- There are any new carbohydrate containing medications
- Constipation is affecting tolerance of the diet
- Meals or enteral feeds are being missed due to fatigue, poor tolerance, vomiting or high ketones
- The correct volume of MCT fat is being given
- The kcal prescription istoo high
- There are a lot of processed foods being used and miscalculated

When the above issues have been addressed but ketones remain low, the ratio of fat to carbohydrate plus protein needs to increase. For example if the patient is following a 3:1 classical KD the ratio should increase to 3.5:1 or 4:1. It is often not possible to increase the ratio much above 4:1 and still provide adequate protein for growth. An alternative option is to replace some LCT fat with MCT fat to improve seizure control. MCT should be increased gradually.

Where the high fat meals are not well tolerated KetoCal 4:1 LQ could be considered for children over 1 year old, as a supplement drink to reduce the fat content ofmeals.

2.9. Phase 4 – Weaning patient from ketogenic diet

A minimum 3 month trial period is undertaken. At the end of this trial period the patient is reviewed in MDT KD clinic by the paediatric KD team. If there has been no improvement in seizure control, the patient will then be weaned slowly from the KD (see appendix 10). Ideally a seizure reduction of ≥50% will be recorded. However, the KD team acknowledges that quantifying seizure activity can often be challenging. Where the parents/ carers feel that benefits to seizure control, behaviour, alertness and/ or other cognitive factors outweigh the burden of KD and the risk of side effects then continuation of KD can be considered. If the KD is seen to have had a positive effect on seizure control, the patient can continue on KD for 2 years, at which point a trial weaning period should be considered in discussion with the family and KD team.

This 2 year period is traditionally based on a similar time period used for anticonvulsant drugs, which are often discontinued after that time in children who become seizure-free [9]. This is understandably a very worrying time for the parents/ carers, particularly if their child is seizure free as a result of KD. They should be reassured that the benefits realised whilst undertaking KD can often be sustained once the diet has been weaned off. Additionally, the child can restart KD if seizure control worsens within a defined period (e.g. 2 months).

Patients with metabolic conditions; GLUT-1 deficiency syndrome or PDH deficiency should continue their KD long term and not trial weaning from KD.

2.10. Management of patients on KD admitted to hospital

Please refer to 'Management of Children on Ketogenic Diet for Epilepsy' Trust Ref C255/2016.

In the absence of a diet kitchen facility to prepare ketogenic meals for inpatients there is a 'Ketogenic/Metabolic Diet Loose Foods Menu' available via the catering department. This is a list of foods from which parents/ carers can choose items to make up a meal. The foods would need to be weighed by the parents/ carers at the bedside in accordance with the most recent KD plan. To calculate appropriate portions (or 'choices') the parent/ carer should use their KD food lists (provided at the start of KD) as they usually would do at home.

Parents/ carers will either bring in their own scales from home or these can be provided temporarily by the Nutrition & Dietetic Service, Ward 27 chef or milk kitchen.

3. Education and Training

Training is available to support the use of this Clinical Guideline via the Nutrition and Dietetic Service. Only appropriately trained Senior Specialist or Senior Paediatric Ketogenic Dietitians should advise a patient on a ketogenic diet.

4. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Patient reported outcome measure (PROMs): Number of Seizures	Patient food and seizure diary and at KD clinic review.	Senior Specialist Paediatric Ketogenic Dietitian	3-6 monthly	Clinic Dit3 letter
PROMs: Number of acute hospital admissions, reason for admission and length of stay	Parent/carer report at KD clinic review.	Senior Specialist Paediatric Ketogenic Dietitian	3-6 monthly	Recorded using dietetic KD clinic proforma (see appendix 11)
PROMs: serum ketone level	Parent/carer report at KD clinic review.	Senior Specialist Paediatric Ketogenic Dietitian	3-6 monthly	Recorded using dietetic KD clinic proforma (see appendix 11)
Clinician rated outcome measure (CROMs): Type and dosage of anti epileptic medication.	KD clinic review.	Senior Specialist Paediatric Ketogenic Dietitian	3-6 monthly	Recorded using dietetic KD clinic proforma (see appendix 11)
CROMs: Anthropometry	KD clinic review. Weight and height/ length measured for all children. Head circumference measured for infants <2 years old.	Senior Specialist Paediatric Ketogenic Dietitian	3-6 monthly	Recorded using dietetic KD clinic proforma (see appendix 11)

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6. Key Words

Ketogenic Diet, Paediatric Epilepsy

CONTACT AND REVIEW DETAILS	
Guideline Lead (Name and Title): Ellen Wilford – Speciality Clinical Lead	**Executive Lead**

Details of Changes made during review:

- Reflect new NICE guidance: addition of reference [11]. Change under referral criteria, no longer need to have trialled 2 anti-eplieptic drugs
- Removed: give weighing scales at pre-assessment appointment
- Added: 'Consent to contact by email form' to pre-assessment appointment starter pack contents
- More general reference to 'electronic ketogenic calculators' rather than specific ones as more options are becoming available.
- Changes to education session starter pack contents to reflect availability of resources and newly developed/updated resources
- Removed MCT fat may have an independent benefit on seizure activity due to contradictory evidence.
- UHL extension telephone numbers updated.
- Added, low carbohydrate foods high in protein as a cause of low ketones due to practical experience
- Reduction in blood and urine tests required in appendix 1
- New starter checklist updated as appendix.
- Updated products list in appendix
- Added reference to extended scope dietitians role
- Updated Title of Guideline Lead

Form 1: Venous blood	Sample Required
3-OH BUTYRATE	From 1 single YELLOW paediatric bottle

Form 2: Venous blood	Sample Required
Lactate	Both samples can be done from 1 single YELLOW paediatric bottle on ICE straight to lab
Glucose	

Form 3: Venous blood	Sample Required
Acylcarnitine (request acylcarnitine and free and total Carnitine will also be reported)	1 orange paediatric lith hep bottle.

Form 4: Venous blood	Sample Required
Vit A, SE Drug LEVEL eg Sodium Valproate if requested by Consultant Neurologist	ALL SAMPLES can be done from 2 BROWN paediatric bottles
Amino Acids **Baseline investigation only**	1 paediatric orange lith hep bottle

Form 5: Venous blood	Sample Required
INR **Baseline investigation only**	1 paediatric green bottle (1.4mls)
FBC	1 paediatric red EDTA bottle
U & E, LFT, Bone Profile, Fasting Chol/trig/HDL Bicarbonate Urate Folate **Baseline investigation only** Ferritin Vit D Vit B12 **Baseline investigation only**	These could all be done out of 2 FULL brown bottles If you have the possibility of getting an extra brown bottle here it would be preferable as vitamin B12 and vitamin D require a substantial sample.

Form 6: Urine	Sample Required - Urine
Calcium:creatinine ratio Amino Acids **Baseline investigation only** Organic Acids **Baseline investigation only**	1 urine bottle

The blood and urinalysis investigations listed above must be undertaken prior to commencing KD. These results are discussed and confirmed with the Consultant Paediatric Neurologist. These investigations must then be undertaken at 3 months on KD and then every 6 months while on KD. For infants <2 years old an additional set of these investigations (not including Vitamin D, lipid profile or free and acyl carnitines) should be undertaken at 6 weeks on KD [10]. The dietetic assistant reminds the parents/ carers when the bloods and urinalysis are due. The Dietitian checks the results on iLab or ICE and reports any abnormal results to the Consultant Paediatric Neurologist, with the suggested treatment if appropriate e.g. increasing the vitamin and mineral preparation in response to low trace element results.

Appendix 2: KD New Starter Checklist

Ketogenic Diet – New patient starter checklist – All to be completed before KD starts

Pre-Assessment

- ☐ Patient demographics added to "KD Service April 16 onwards" spread sheet
- ☐ Information pack provided at pre-assessment appointment
- ☐ Includes scales

Screening

- ☐ Foods trial questionnaire returned
- ☐ Weighed 5 day food & seizure diary returned
- ☐ Food diary analysed on diet plan
- ☐ Send 'Guide to Classical KD' or 'Guide to MKD' leaflet by email
- ☐ Biochemistry done
  - ☐ Blood/urine forms x 7 complete and given to parents
  - ☐ Bloods ☐ Urine
  - ☐ Repeats biochemistry completed or not needed
- ☐ ECG in infants <2 years old and patients with a history of heart disease

Medications

- ☐ Medications cho content reviewed via medicines info.
- ☐ Medications changed to reduce cho if necessary.

Education

- ☐ Ask Natalie to book a room for education session and send parents an invitation letter.
- ☐ Book neuro nurse for ketone meter education

Prescriptions

- ☐ Calculate individual KD prescription
- ☐ Calculate appropriate dose of Fruitivits / walnut oil / liquigen as appropriate
- ☐ Meal plans formulated
- ☐ Request GP prescription for products e.g. Fruitivits, KetoCal, Liquigen, dextrogel
- ☐ Neuro consultant advised on low cho paracetamol & ibuprofen option

☐ Patient registered with Nutricia Homeward if necessary

- ☐ Parent consent obtained for Nutricia Homeward registration
- ☐ Switch child to Nutricia Homeward Metabolics if already on Homeward. Call metabolics and they will do this straight away.

☐ Parents/carers attend education session

- ☐ Illness including hyperketosis & hypoglycaemia explained
- ☐ Education on ketone & BM monitoring with Neuro Nurse received
- ☐ Food choices lists provided and KD meal worksheets completed
- ☐ Hopes & expectations questionnaire returned
- ☐ Parents/Carers advised to inform school/nursery/respite/ any informal childcare providers of plan to start KD and plan for managing hyperketosis/hypoglycaemia
- ☐ Discuss parents role with providing schools/nursery/ respite centres with recipes/ plan for school meals or packed lunches
- ☐ MF & Nutricia starter packs provided
- ☐ EKM demonstration provided if on classical KD

- ☐ Give 'I am on KD' emergency admission card
- ☐ Provide details to request Daisy Garland welcome box
- ☐ Give a urine collection kit for the 3 month biochemistry

☐ Invite school / nursery /respite centre to an education session or place on waiting list

Deliveries

☐ Patient received 1 month supply of nutritional prescription products

☐ Patient received 1 month supply of ketone/BM strips and lancets received via GP prescription

☐ If under 1yr old arrange admission to start KD

Admin

☐ Baseline outcome measures (from pre-assessment appointment) added to "outcome measures" spread sheet

☐ Hazard alert for medical notes done

☐ Update "KD service April 16 onwards" spread sheet with start date and add patient to "clinic rv and new" and "bloods & food diaries reminder" spread sheets for 3 month up to 2 years on KD clinic appointment.

The Dietitian decides on the most suitable KD for the patient. This will be influenced by the route of feeding available and the age of the child. Those orally feeding generally start on the Modified KD. Those enterally tube fed generally start on the Classical ketogenic diet because prescribed ketogenic enteral tube feeds are available using the classical calculation method. It is recommended that children under 2 years old follow a classical ketogenic diet regardless of route of feeding [10]. Parents/ carers opinions are welcome if appropriate (e.g. the Low Glycaemic Index diet is not offered at UHL).

CLASSICAL KETOGENIC DIET

The classical KD is calculated in a ratio of grams of fat to grams of protein plus carbohydrate. It is the strictest of all of the KDs, in that it allows the least amount of carbohydrates. It is standard practise to start at a ratio as low as 2:1 and build up towards a 4:1 ratio depending on ketosis. Younger patients may achieve adequate ketosis on lower ratios. LCT is the main fat source e.g. butter, cream, oil and mayonnaise.

The patient is advised to have a certain number of meals +/- snacks; all of which must be individually calculated and in the same ratio. Specialist enteral feeds are available that can be altered to suit the dietary ratio and prescription e.g. KetoCal 4:1 LQ (4:1 vanilla or unflavoured, Nutricia), KetoCal Powder (3:1 unflavoured, 4:1 unflavoured and vanilla, Nutricia), KetoCal 2.5:1 MCT Multifibre LQ (Vanilla, Nutricia), and Ketovie 4:1 (Cambrooke) see appendix 4 for detailed information regarding enteral feeds. See appendix 5 for an example of calculating this diet.

In infants, the classical KD is the recommended choice commencing with a 1:1 ratio as an inpatient and building up towards a 3:1 ratio [10].

Where an oral diet is required 1 fat choice = 5g fat, 1 protein choice = 3g protein and carbohydrate will be prescribed in grams.

MEDIUM CHAIN TRIGLYCERIDE (MCT) DIET

This diet has not been used in UHL since 2018 due to a more complex method of calculating protein choices and the change in practice towards using MCT fats in higher doses in other ketogenic diets. The MCT KD has up to 60% of energy derived from MCT fat. Medium chain triglycerides yield more ketones per kcal of energy than LCT. This increased ketone potential allows less total fat to be used, and slightly more protein and carbohydrate than the classical KD. However, MCT fat can cause gastrointestinal discomfort in some children with reports of abdominal cramps, diarrhoea and vomiting, increasing the MCT fat gradually may prevent these side effects. MCT fat can be given in the diet as a fat emulsion (Liquigen, SHS) or oil (MCT oil, SHS). The MCT fat must be taken at every meal and snack, either incorporated into the meal or taken as a drink. This diet is calculated in choices (previously known as ‘exchanges’) of fat, protein and carbohydrates. The patient is prescribed a number of choices for each and can divide these as they wish throughout each day, ideally spreading the choices

equally. Both the classical KD and MCT KD are comparable in terms of efficacy (seizure reduction) and tolerability [17].

Typically we start at 45% of energy from MCT initially. Fine tuning the diet for optimal ketosis usually results in an MCT level of 45-55%. It has generally been well tolerated with good compliance. See appendix 5 for an example of calculating this diet.

1 fat choice = 5g LCT fat

1 protein choice = 6g protein + 6g LCT fat Carbohydrate will be prescribed in grams

MODIFIED KETOGENIC DIET

This KD has gained a lot of popularity amongst UK centres in recent years. A similar ketogenic diet in the USA is known as the 'modified atkins diet'. A publication by the Ketogenic Dietitians Research Network (KDRN) described the use of this diet in the uk [20]. Carbohydrate is restricted to 10-30g daily so generally a lower allowance than on the MCT KD. A recommended number of fat choices are prescribed at UHL, although not all centres in the UK give specific advice on the quantity of fat to consume. Some centres simply encourage high fat foods at every meal. Quantified advice on fat is given at UHL to support parents to understand the very large amounts of fat required and to facilitate effective manipulation of the diet when required. Once modified KD is established it may be appropriate to stop quantified prescriptions of fat and encourage a high fat intake at each meal providing the child has stable ketosis; the parents/carers are confident with estimating the quantity of fat required and that the ketogenic diet team are confident in the parent/ carer ability to do this. MCT fats can be incorporated into the modified KD (by prescribing quantified amounts) to increase ketones and improve seizure control. Protein foods which are not sources of carbohydrate (e.g. unprocessed beef, chicken or eggs) are not restricted and do not have to be weighed or measured. There has been good success reported with this diet in adolescents and adults due to its less restrictive nature [21, 22]. See appendix 7 for an example of calculating this diet.

1 fat choice = 5g fat, protein choices are not used as protein is allowed freely and carbohydrate will be prescribed in grams.

THE LOW GLYCEMIC INDEX TREATMENT (LGIT)

This KD focuses particularly on the glycaemic index (GI) and type of carbohydrate. The GI is a measure of the effect of carbohydrates on blood glucose levels. When carbohydrates are digested, they release glucose into the bloodstream. Carbohydrates that digest rapidly have a high GI. Carbohydrates that are digested slowly have a low GI. Foods are rated based on their GI values ranging from 0 to 100. The LGIT includes foods that have a GI of 50 or lower. In addition to the GI, the digestion of a carbohydrate food is slowed by foods that are eaten at the same time that contain either fat or fibre. Therefore, meals are balanced with sources of fat, protein and a low glycaemic index carbohydrate. This diet is not used often in the UK and is not offered at UHL.

These are the most common products used while on KD; however it is not an exhaustive list. It will not include any new products brought to market between the guideline development and review dates. All products below can be prescribed by the patients GP, however KetoCal 3:1 and KetoCal 4:1 powder and KetoCal 4:1 and KetoCal 2.5:1 MCT Multifibre liquid can only be dispensed via the Nutricia Homeward delivery service.

Parents/ carers' should be advised to bring all their child's usual medications and feed constituents when being admitted to hospital, in case of low hospital stock levels.

Liquigen (Nutricia) is a medium chain triglyceride (MCT) fat emulsion consisting of 50% MCT oil and water. 2ml of Liquigen = 1g fat. The recommended dose can be taken as a 'shot' from a syringe, added to semi skimmed/ skimmed milk or diet soft drinks, added to an enteral feed or added during food preparation e.g. mashed potato, cauliflower cheese.

MCT Oil (Nutricia) is a liquid containing pure medium chain triglycerides. 1ml = 1g of fat. It can be used in cooking but should not be used to 'deep fry' foods as it has a higher flash point than other oils. It can also be incorporated into enteral feeds.

KetoCal 2.5:1 MCT Multifibre LQ (Nutricia) is a nutritionally complete 2.5:1 ready-made liquid feed in vanilla flavor that can be taken orally or administered via an enteral feeding tube for children from 8 years old.

KetoCal 3:1 (Nutricia) is a nutritionally complete powdered feed based on the classical KD. It can be prepared and taken orally, or delivered via a nasogastric/gastrostomy tube. Ketocal 3:1 is suitable for use from birth to six years of age. The standard ratio is 3:1 i.e. 3g fat to every 1g of protein plus carbohydrate. The ratio can be lowered using a carbohydrate and/ or protein powder (e.g. Polycal and Protifar).

KetoCal 4:1 (Nutricia) is a nutritionally complete powdered feed, based on the classical KD. It can be prepared and taken orally, or delivered via a nasogastric/ gastrostomy feeding tube. Ketocal 4:1 is suitable for children aged 1 year and above. The standard ratio is 4:1 i.e. 4g fat to every 1g of protein plus carbohydrate. The ratio can be lowered using a carbohydrate and/or protein powder
(e.g. Polycal and Protifar). It is standard practice at University Hospitals of Leicester to begin at a 1:1 ratio increasing gradually as necessary. Ketocal 4:1 is available in vanilla and unflavoured.

KetoCal 4:1 LQ (Nutricia) is a ready-made liquid version of the 4:1 ketocal powder. Available in vanilla* and unflavoured.

***Ketovie (Cambrooke)** A nutritionally complete ready-made liquid feed at a 4:1 ketogenic ratio. Available in vanilla or chocolate flavour. This product contains docosahexaenoic acid, fibre and carnitine.

***Ketovie Peptide (Cambrooke)** A nutritionally complete ready-made liquid feed at a 4:1 ketogenic ratio. The only extensively hydrolysed whey protein ketogenic formula available. Contains MCT fats.

Calogen Neutral (Nutricia) is a fat emulsion consisting of 50% long chain triglycerides (LCT). 2ml=1g fat. It can be used as a substitute for double cream, particularly if the

patient is cow's milk intolerant or if their cholesterol level is increasing. Calogen can be added to enteral or modular feeds as a source of LCT.

***Betaquick (Vitaflo)** is a MCT fat emulsion consisting of 20% MCT with sweetener. 5ml = 1g fat. The recommended dose can be taken as a 'shot' from a syringe, added to semi skimmed/ skimmed milk or diet cola, added to an enteral feed or added during food preparation e.g. mashed potato, cauliflower cheese.

Protifar (Nutricia) is a protein powder. It can be used in modular ketogenic enteral feeds, emergency milkshakes, or added to Ketocal to decrease the fat ratio. It is a useful product for patients who are part enterally fed and part orally fed who may rely on soft easy textures e.g. fruit and cream.

Polycal (Nutricia) is a powdered carbohydrate supplement which can be used as a treatment for hyperketosis/ hypoglycaemia. It can also be added to modular ketogenic enteral feeds, emergency milkshakes or Ketocal. 1g of Polycal = 1g carbohydrate.

***MCT Procal (Vitaflo)** is a neutral flavoured powder for food fortification. It is suitable from 1 year of age. Each sachet contains 10g MCT fat, 3.3g carbohydrate and 2g protein. It is the only non- liquid form of MCT fat available on prescription and may be useful when compliance with the MCT KD is compromised due to a refusal to drink MCT oil/emulsion and/or the soft texture of foods when MCT fat has been added as a liquid.

***K.yo (Vitaflo)** is a ready to eat semi-solid pudding style supplement. It is nutritionally complete for children up to 10 years old. Available in chocolate flavour. This supplement can be useful to aide compliance to the diet where taste fatigue is a problem.

***3:1 Classic Bar (Ketocare)** is a ready to eat nut bar is a 3:1 classical ketogenic diet ratio. The bar can be used on the MCT KD or Modified KD also. The bar can be useful as a snack or as part of a meal (e.g. packed lunch) when resources for food preparation are limited. The bar is also high in fibre but low in carbohydrate due to the inulin content. Ketogenic diets are often low in fibre so this can be a useful addition to help manage constipation.

***3:1 Classic Chicken (Ketocare)** is a chicken soup meal in a 3:1 classical ketogenic diet ratio. The meals are available in 135g pouches. The meals can be adapted to other ketogenic ratios by either adding carbohydrate (e.g. bread) or fat (e.g. double cream). The meal can be used on MCT KD or Modified KD also.

FruitiVits (Vitaflo) is the vitamin and mineral supplement of choice due to superior palatability. It is a carbohydrate free orange flavoured powder in 6g sachets. Careful comparison of the product with the patient's vitamin and mineral requirements is recommended. Younger children might require only 2-3g daily. Any remaining should be sealed in an airtight container for use the following day. The dose may need adjusting in response to abnormal blood results and as the child ages. Additional vitamin D and iron supplementation is often needed in response to low serum levels. Do not greatly increase the FruitiVits in order to improve the vitamin D or iron levels, this may result in toxicity (e.g. vitamin A) and be ineffectual at treating the Vitamin D or

iron deficiency. Additional electrolytes e.g. sodium chloride and potassium chloride will need to be added to modular feeds and possibly to Ketocal to meet the patient's requirements.

***Phlexy-Vits (Nutricia)** is a carbohydrate free vitamin and mineral supplement available in powder and tablet form but are only ACBS approved for aged 11 years and over. They can be used in younger children but with caution particularly when choosing a suitable dose. It does not contain choline unlike fruitivits.

Resource Optifibre (Nestle) is a powdered soluble dietary fibre that can be used for those patients suffering from constipation. However it does contain carbohydrate (0.3g per 5g scoop) and this should be accounted for in the calculation of the KD prescription.

***Thixo-D Cal-Free (Ecogreen Technologies Ltd)** is a kcal and carbohydrate free thickener that can be used in conjunction with a KD. This is not ACBS approved however there is no other carbohydrate free thickener available so the GP can prescribe this at their discretion. It is not a stock item at UHL Pharmacy; however it can be ordered for inpatients.

* These products are not stocked at University Hospitals of Leicester NHS Trust.

The ratio should be increased gradually and it may be necessary to increase in 0.5:1 or 1:1 increments, depending on the patient. The patient would remain at each stage for 1-3 days depending on their response and level of ketosis. Ketocal Liquid or Ketocal powder can be used as a supplementary drink/ enteral feed or as a sole source of nutrition in those completely enterally fed. The standard ratio of the Ketocal liquid and powder is 4:1; however Polycal and Protifar can be added to decrease the ratio. Ketocal 3:1 powder is also available as an infant formula and more recently KetoCal 2.5:1 liquid has been made available.

When commencing the classical KD, orange juice or a similar carbohydrate containing drink can be used to reduce the ratio easily, without having to calculate a full set of meal plans at each individual ratio. The orange juice would be reduced with each increasing 0.5:1 or 1:1 increment. Other centres have had good success commencing with one ketogenic meal on day 1, two ketogenic meals on day 3-4 and so on until on full diet.

CALCULATION OF ENERGY AND PROTEIN REQUIREMENTS

5 year old girl, 18kg, weight and height on 50th centile Protein requirement = 1g/kg/day = *18g/day*
EAR for energy 82kcal/kg = 1476kcals

Energy prescription = 80% of EAR for energy as she is immobile and wheelchair bound = *1180kcals*

Calculation of energy requirement should also take into consideration current energy intake (from enteral tube feeding regimen and/ or 5 day weighed food diary) and the associated growth trajectory, if this information is available.

INTRODUCTION OF DIET AT 1:1 RATIO

Number of dietary units

Each diet unit = 1g fat (9 kcals):1g protein + carbohydrate (4 kcals) = 13kcals 1180/13 = 91 dietary units

Quantity of fat = 91 x 1 = 91g/day
Quantity of protein + carbohydrate = 1 x 91 = 91g/day
Quantity of protein at 1g/kg/day = 18g/day
Quantity of carbohydrate = 91-18 = 73g/day (nb if medications contain cho; it needs to be accounted for so the daily carbohydrate allowance will be lower)

Daily prescription 1:1 ratio

91g fat
18g protein
73g carbohydrate
Divided equally among 3-4 meals

4 meals/feeds per day will include:

22.75g of fat (round to 23g fat)
4.5g protein (round to 5g protein)
18.25g carbohydrate (round to 18g carbohydrate)

For oral intake this could be expressed as 5 fat choices, 1 ½ protein choices and 18g carbohydrate per meal

INCREASING RATIO TO ACHIEVE ADEQUATE KETOSIS

2:1 RATIO

Number of dietary units
Each diet unit = 2g fat:1g protein + carbohydrate = 22kcals
1180/22 = 54 dietary units

Quantity of fat = 54 x 2 = 108g/day
Quantity of protein + carbohydrate = 1 x 54 = 54g/day
Quantity of protein at 1g/kg/day = 18g/day
Quantity of carbohydrate = 54-18 = 36g/day (nb if medications contain cho; it needs to be accounted for so the daily carbohydrate allowance will be lower)

Daily prescription 2:1 ratio

108g fat
18g protein
36g carbohydrate

Divided equally among 3-4 meals

4 meals/feeds per day will include

27g of fat
4.5g protein (could be rounded to 5g)
9g carbohydrate

3:1 RATIO

Number of dietary units

Each diet unit = 3g fat : 1g protein + carbohydrate = 31kcals
1180/31 = 38 dietary units

Quantity of fat = 38 x 3 = 114g/day

Quantity of protein + carbohydrate = 1 x 38 = 38g/day
Quantity of protein at 1g/kg/day = 18g/day
Quantity of carbohydrate = 38-18 = 20g/day (n.b.if medications contain cho; it needs to be accounted for so the daily carbohydrate allowance will be lower)

Daily prescription 3:1 ratio

114g fat
18g protein
20g carbohydrate

Divided equally among 3-4 meals

4 meals/feeds per day will include

28.5g of fat (round to 29g)
4.5g protein (round to 5g protein)
5g carbohydrate

4:1 RATIO

Number of dietary units

Each diet unit = 4g fat : 1g protein + carbohydrate = 40 kcals
1180/40 = 29.5 dietary units

Quantity of fat = 29.5 x 4 = 118g/day
Quantity of protein + carbohydrate = 1 x 29.5 = 29.5g/day
Quantity of protein at 1g/kg/day = 18g/day
Quantity of carbohydrate = 38-18 = 11.5g/day (nb if medications contain cho; it needs to be accounted for so the daily carbohydrate allowance will be lower)

Daily prescription 4:1 ratio

118g fat
18g protein
11.5g carbohydrate

Divided equally among 3-4 meals

4 meals/feeds per day will include

29.9g of fat (round to 30g)
4.5g protein (round to 5g)
2.9g carbohydrate (round to 3g)

For oral intake this could be expressed as 6 fat choices, 1 ½ protein choices and 3g carbohydrate per meal.

EXAMPLE ENTERAL MEAL PLAN AT 2:1 RATIO

Classical Ketogenic Diet Plan Calculations

Food	Fat (g)	CHO (g)	Protein (g)	
590ml Ketocal 4:1 Liquid	87.3	3.6	18.2	
40ml Calogen Neutral	20			
33g Polycal		31.7		
				Ratio
Total	107.3	35.3	18.2	107.3/53.5 2:1
Kcals	965.7	141.1	72.8	1180

Recipe

590ml Ketocal
40ml Calogen
33g Polycal

Mix ingredients and add water up to 700ml
5 equal feeds per day of 140ml

Total = 700ml

Fluid requirement = 1400ml, additional 700ml as fluid flushes needed throughout the day

It is now more common for the paediatric ketogenic dietitians to calculate classical KD enteral feed regimens using Microsoft Excel spreadsheets than manually as above.

EXAMPLE ORAL MEAL PLAN AT 2:1 RATIO

Breakfast
9g Weetabix
14ml double cream
125ml Ketocal liquid
Mix the double cream and Ketocal , use enough to mix with Weetabix and remainder as a drink

Mid-Morning
Sugar free squash

Lunch
10g cooked beef mince
15g carrots
10g onion
5g garlic
10g olive oil
26g double cream
15g cooked spaghetti

Mid-Afternoon
Sugar free jelly

Evening Meal
13g cooked chicken breast
20g butter
16ml double cream
44g boiled potato
30g swede

Bedtime shake
150ml Ketocal liquid
8ml double cream
8g Polycal powder

CALCULATION OF ENERGY AND PROTEIN REQUIREMENTS

Active 11yr old girl

32kg 1600kcal/day (av. of EAR + food diary)

RNI pro = 41.2g/day Add 30g to allow for extra eaten

Estimated protein intake = 71g/day (284kcal/day) compare this against weighed food diary analysis

CALCULATION OF DIETARY PRESCRIPTION

Decide on the initial level of carbohydrate restriction between 10-30g/day [20, 23]. This is dependent on the age and size of the child and can also be affected by non-dietary sources of carbohydrate e.g. medication. The carbohydrate restriction is usually lower initially then increased as ketosis allows. The starting carbohydrate restriction should be decided by a Senior Specialist Paediatric Dietitian with experience in ketogenic diet.

Carbohydrate restriction = 15g/day (60kcal/day)
LCT Fat prescription
Calculate kcal intake from protein and carbohydrate: 284 + 60 = 344kcal
Total kcal – kcal from protein and carbohydrate: 1600 – 344 = 1256kcal remaining
Calculate grams of fat: 1256/9 = 140g

Daily prescription:

15g carbohydrate (e.g. 3 x 5g)
140g LCT fat (e.g. 28 x 5g choices)
Aim for approximately 1600kcal/day and 41 - 71g protein per day on first ketogenic food diary analysis.

Frutivits 3g - Use a Nutritional analysis programme such as Nutritics to guide appropriate dosage of FruitiVits and electrolytes to meet at least the LRNI but ideally RNI for all micronutrients.

ORAL MEAL PLAN EXAMPLE

Meal	Recipe	
Breakfast	8g Weetabix 21g ground brazil nuts 40ml double cream Use water, Alpro soya unsweetened milk or Alpro unsweetened almond milk to thin down the cream. Or drink any excess cream	5g cho 7 fat choices
Lunch	Roast lamb, chicken, pork or beef 80g boiled celeriac mashed 60g boiled carrots 35g butter Mix some butter into the mash and the rest on the carrots	5g cho 7 fat choices
Dinner	13g wholemeal bread toasted Bacon Chicken 60g avocado 10ml olive oil 5g butter 20g mayonnaise Fry the chicken and bacon in the oil (serve any oil in the pan onto the plate). Spread the butter on the toast. Mix the bacon and chicken in the mayonnaise. Serve on the toast with the avocado.	5g cho 7 fat choices
1hr before bed	70ml Double cream Crusha chocolate no added sugar Use water, Alpro soya light (fresh) milk or Alpro unsweetened almond milk to thin down the cream.	7 fat choices

- Calculate an individual dietary prescription and meal plan as previously discussed

- Liaise with Pharmacy to arrange prescription and supply of necessary products. For example Ketone testing strips, Liquigen and FruitiVits. Ensure Pharmacy have adequate supply of these items in stock for the entire inpatient admission and to provide a supply on discharge. For patients who are enterally tube fed, appropriate supply of ketogenic formula e.g. KetoCal or other recipe constituents should be ensured

- Liaise with the Paediatric Pharmacist regarding the carbohydrate content of the patient's medications. Make the medical team aware that carbohydrate content of drugs should be considered if prescribing any new medications

- A 'Loose Foods Menu' of basic meal ingredients is available from catering. This can be used for parents/ carers to order foods so that they can then weigh carbohydrate, protein and fat sources at the bedside. If parents/ carers are not available or able to weigh foods then request that this is done via the catering department and provide detailed meal plans with weights not choices. Usually the catering department will request the ward 27 chefs take on this role. Inform the catering manager at the start of an admission (or in advance if possible) that a patient on a ketogenic diet has been admitted

- Often special catering provisions are required (e.g. involvement of ward 27 chefs, special purchase of non-stocked food items etc). If required, non stock items such as double cream, strawberries, suitable yoghurts, snacks and sugar free jelly should be purchased by catering and supplied to the ward. Catering will not modify the texture by puréeing the meal. If puree meals are necessary the Dietitian will have to investigate the suitability of the pre-packed puree meals. These are usually reformed in their constituent parts so the vegetables (cho) meat (protein) and potatoes (cho) could be weighed and additional butter or oil (fat) added

- If enteral feeds are to be prepared, inform the milk kitchen and provide a recipe. Ext. 16465

- Prepare an individual patient care plan to guide nursing staff on the appropriate management of the patient. The care plan should cover: monitoring, how to treat hyperketosis or hypoglycaemia, and feed plan (oral & or enteral). Include a copy of *'Management of Children on Ketogenic Diet for Epilepsy'* Trust Ref C255/2016 which includes guidance on choice of IV fluids if required

- If the child is enterally tube fed prescribe polycal 'as required' in appropriate doses for the treatment of hypoglycaemia and separately for hyperketosis with instructions that it should be dissolved in 30-50ml water

- Ensure an interpreter is booked to undertake education sessions with the family if English is not their first language

- Ensure you are available to provide intensive support and education for the family while admitted, providing the usual information to parents as described in Stage 3 – Initiating and maintaining ketogenic diet

Ensure a hazard sticker and alert form is placed in the patient's medical notes.

- The ketone meter used by nursing staff for inpatients in UHL is not available for community use. UHL does not stock ketone and glucose strips that would be appropriate to supply on discharge for use at home. Therefore the paediatric neurology nurse specialist should request a prescription from the GP for ketone and glucose strips, lancets and a sharps bin at the earliest opportunity. The paediatric neurology nurse specialist can also request free sample supplies of strips and lancets to be given on discharge however availability can be variable

Serum Ketones aim 2.0 – 5.0mmol/l

It is preferable to use serum ketone testing for monitoring because it is more accurate than urine testing; urine is not always available on demand because children cannot always provide a fresh sample when required; urine will indicate the ketone level an hour or more ago and urine test strips cannot indicate hyperketosis (urinary ketones >16mmol/l). However, in exceptional circumstances where KD is already established and proved effective and parents are strongly against daily serum testing then providing that the consultant neurologist agrees to urine ketone testing they could switch to urinary testing. This should only be on the understanding that serum ketone testing should still be used when necessary e.g. when an adequate urine sample cannot be obtained or when urine ketones are high and serum ketone testing is needed to identify and potentially manage hyperketosis. There is a ketone breath scanner called 'ketoscan mini' available to market which could be considered but has not been used by the UHL paediatric ketogenic diet service to date. Children would have to comply with breath control instructions to use the device.

Ketones should be checked twice daily, for example before breakfast and before bed time but testing may need to increase to 4 times daily or more in times of illness or when closer monitoring is required. Ketones should ideally be checked before meals to get a true reading of the ketone levels. However, if checking urinary ketones it will be near impossible to dictate the timings of urination and ketone testing for example, in young children or children with developmentaldelay.

Some patients can maintain good seizure control on lower ketones but the majority of patients need to be in the higher ketone range. If the patient continues to experience seizures and has lower ketones the diet should be fine tuned further to optimise ketosis. If the patient has very erratic ketones, it may be beneficial to check the ketones 3-4 times daily in order to identify when the ketones are lower and if this is affecting seizure control. Ketones are generally lower in the morning after the overnight fast.

If it has been identified that seizure control is best when serum ketones approach or exceed 5.0mmol/l then it may be appropriate to review the upper threshold for ketone level to 5.5mmol/l or 6.0mmol/l on an individual patient basis. This decision is made by the SeniorSpecialist Dietitian with extended scope role approved or the Consultant Paediatric Neurologist.This information should be documented on individual care plans and in a letter to parents/carers and the MDT.

Serum blood glucose

Blood glucose should be maintained above 2.5mmol/l. It is unusual for patients to experience hypoglycaemic incidents so blood glucose is not routinely tested. We test only in the first few weeks of KD and during illness.

High Ketones (Hyperketosis) – higher than 5.0mmol/L

Ketones can increase unexpectedly when a child is unwell or a recent change has been made to the diet. Your child's blood ketones may be greater than 5.0mmol/L.

Symptoms

- Ketones above 5.0mmol/l
- Lethargy
- Fatigue
- Irritability
- Rapid breathing / panting
- Increased heart rate
- Facial flushing
- Unexpected vomiting

If you suspect your child is experiencing the above symptoms;

- Check your child's ketone levels
- Check your child's blood glucose levels

It rarely happens. However, if your child is experiencing the above symptoms and/or serum ketones are high - the body needs some sugar to reduce the ketones back into the ideal range. Sugar is used because it's absorbed more quickly than starchy carbohydrates such as bread. Guidance on treatment is below. Telephone your Ketogenic diet team if you have any questions or worries regarding this.

Treatment

Give one of the following to provide 5g carbohydrate from sugar;

- 50ml pure fruit juice
- 50ml fizzy drink (Cola or lemonade – NOT 'diet'/'zero' varieties)
- 2 teaspoons of jam
- 1 teaspoon of sugar
- (NGT/PEG treatment) - 1 level scoop Polycal powder mixed with 30ml water

1) Recheck the ketone levels in 20 minutes, if they have not decreased to under 5.0mmol/L then **treat again**

2) Recheck ketone levels in 20 minutes and if they have not decreased to under 5.0mmol/L **treat again and seek advice from your Dietitian, local Doctor or go to your nearest Accident and Emergency department.**

3) Your child may need to come to the hospital. It is very rare that this occurs. While seeking help **continue to check the ketones and treat every 20 minutes** as necessary on the way to hospital.

If you are in any doubt as to whether you should treat or not, please be safe and treat as above. The consequence will be lower ketones for a time but you and your Dietitian can work together to resolve this.

Low Ketones – less than 2.0mmol/l – no action

It is not uncommon for ketones to be lower in the morning after the overnight fast without any fat. If the ketones are under 2.0mmol/l, this does not require any immediate action. Ideally we would adjust the ketogenic diet recipe to optimise the ketones consistently above 2.0mmol/l but this is not always possible. Inform your Dietitian if the ketones are persistently lower than 2.0mmol/l.

Low Blood Glucose (Hypoglycaemia) – less than 2.5mmol/L

Children on ketogenic diets often have lower blood sugar levels than children on a normal non ketogenic diet. This is not a problem unless it drops below 2.5mmol/l and/or symptoms develop. This occurs so rarely that we do not routinely check blood sugar levels at home, only when we commence the ketogenic diet and if your child is unwell and/or admitted to hospital.

Symptoms

- Blood glucose less than 2.5mmol/l
- Lethargy/ Fatigue
- Irritability
- Sweating / Becoming cold and clammy
- Dizziness
- Jittery
- Confused
- Aggressive behaviour
- Pallor / pale appearance

If you suspect your child is experiencing the above symptoms;

- Check your child's blood glucose levels
- Check your child's ketone levels

Both high ketones and low blood sugar levels are treated with carbohydrate.

- If the blood glucose is 2.5mmol/l or above and the ketones are under 5.0mmol/L there is likely another reason for your child's symptoms and he/she does not require treatment with extra carbohydrate
- If blood glucose is under 2.5mmol/l treat immediately with one of the following options to give 10g carbohydrate from sugar
    - 100ml of pure fruit juice
    - 100ml fizzy drink (Cola or lemonade – NOT 'diet'/'zero' varieties)
    - 4 teaspoons jam
    - 2 teaspoons sugar
    - (NGT/PEG treatment) - 2 level scoops Polycal powder dissolved in 50ml water

Note that this is double the amount of treatment that we use to treat high ketones.

1) Recheck blood glucose in 20 minutes and if it has not increased to above 2.5mmol/l; **treat again and call 999 for an ambulance**. Your child may need to be admitted to hospital for an intravenous glucose infusion.
2) While seeking help, **continue to check the blood glucose level's every 20 minutes** and treat as above until the reading increases to above 2.5 mmol/l.

If you are in any doubt as to whether you should treat or not, please be safe and treat

as above.

WEANING FROM ORAL KD

Weaning from KD is very patient specific, these are suggestions only.

If the patient has been seizure free for 2-3 years or more on KD then a weaning period of 2-3 months may be most appropriate, slowly decreasing MCT and LCT fat and slowly increasing carbohydrate foods.

If the patient has been on KD for 2-3 years with some improvement in seizure control then wean over 4-6 week period.

If the patients seizure activity has been unaffected by KD and they are being weaned following a short trial on KD then a 2 week wean from KD would be appropriate.

Changes could be made daily or weekly as appropriate. Once ketones are regularly less than 1.0mmol/l with no undesirable effect on seizure control then the child should have a 'normal' unrestricted diet within days.

WEANING FROM CLASSICAL KD

This will be very patient specific. If the patient is on a 4:1 ratio, it would be acceptable to reduce by 1:1 ratio every 2 weeks, taking 4-8 weeks to wean from KD. However if the patient was highly ketotic on a 1.5:1 or 2:1 ratio, it would be best to reduce the ratio by only 0.5:1 increments every 2 weeks depending on how sensitive the patient is to change.

Protifar and Polycal should be used in the early stages to save parents/ carers having to recalculate new meals at every weaning stage. The Polycal and Protifar can be easily added to meals or shakes to decrease the ratio of fat to protein plus carbohydrate. If the child drinks orally, orange juice could be used in place of Polycal.

Sweet foods can be introduced slowly, once a 'normal' diet is established, i.e. the patient is eating similar meals to the rest of the family at portions that satisfy him/ her.

University Hospitals of Leicester NHS
NHS Trust

Paediatric Ketogenic Diet Clinic

Name:	
DOB:	
NHS:	
S no:	

Date & time:	
Age:	
Weight:	Centile:
Height:	Centile:
Started KD:	
Length on KD:	
Ketones:	

Diet Ratio/Plan & tolerance:

Food & Seizure diary returned? ☐

Biochemistry:

Side effects of KD / Complications / Non AED medications:

Renal USS (if 1yr r/v) ☐

Other:

To Do:

Review:

Signed: **Print:** **Designation:**

1

Parent Reported Outcome Measures

Seizures

1) Type, Frequency & Duration

2) Type, Frequency & Duration

3) Type, Frequency & Duration

Hopes & Expectations

0 = quality of life is poor. 10 = quality of life is very good

How does living with epilepsy and seizures affect your family's quality of life currently?

1) How would you rate where you are regarding this aspect of your life?

2) How would you rate where you are regarding this aspect of your life?

3) How would you rate where you are regarding this aspect of your life?

Clinical Reported Outcome Measures

Anti-Epileptic Drugs (include changes this OPA)

1) AED & dose

2) AED & dose

3) AED & dose

Admissions since last appointment (for any reason)

1) Date of admission

Length of stay

Reason for admission

2) Date of admission

Length of stay

Reason for admission

2