An introductory guide for the practical implementation of the **ketogenic diet (KD)** for the dietary management of epilepsy and neurometabolic disease
Introduction

Purpose
This practical guide is for use as an aid to implementation of the ketogenic diet (KD) for the dietary management of children aged 1 year or over and adults with epilepsy or an inherited neurometabolic disease, i.e. Glut-1 deficiency syndrome (Glut-1 DS) or pyruvate dehydrogenase deficiency (PDHD). The KD can also be used in the dietary management of other conditions including cancer and neurological diseases, however, this guide will focus on dietary management of epilepsy, Glut-1 DS or PDHD only.

Introducing and adjusting the KD is dependent on the individual patient. Practical examples are given in this guide. However, it is the responsibility of the managing health care professional to use clinical judgement to introduce and adjust the KD in the most appropriate way for individual patients, and it may not always be appropriate to follow the examples in this practical guide.

Intended users
This practical guide is:
• for use by health care professionals working with children aged 1 year or over and adults diagnosed with epilepsy, Glut-1 DS or PDHD.
• not for use by parents/caregivers of patients with epilepsy, Glut-1 DS or PDHD, or patients themselves.
• for general information only and must not be used as a substitute for professional medical advice.

Important note
For infants i.e., those under 12 months of age, further guidance from specialist resources should be sought.

Product information
Any product information contained in this practical guide, although accurate at the time of publication, is subject to change. For the most up-to-date detail, including allergen content, refer to product labels and/or www.vitafloweb.com.

Disclaimer
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This practical guide should be read in conjunction with local, national and international guidelines and best practice for the dietary management of epilepsy, Glut-1 DS or PDHD. Information contained within the guide is based on the most recent scientific evidence available on the use of the KD in the dietary management of epilepsy, Glut-1 DS and PDHD at the time of publication.

This practical guide does not establish or specify particular standards of medical care for the treatment of any conditions referred to in this practical guide.

Vitaflo (International) Ltd does not recommend or endorse any specific tests, procedures, opinions, clinicians or other information that may be included or referenced in this practical guide.

It is the responsibility of the Multi-Disciplinary clinical Team (MDT), i.e. a dedicated keto-team*, to ensure patients managed on the KD are suitable to undergo this form of dietary therapy and they undertake and implement all the assessments, procedures, investigations and monitoring required in accordance with locally agreed procedures specific to the intervention.

* N.B. The term ‘keto-team’ is a generic description for those healthcare professionals (for example, dietitians, clinicians, nurses) involved in the implementation, follow-up and care of patients on a KD.

Abbreviations

AED Anti-epileptic drug
CKD Classical ketogenic diet
DRE Drug-resistant epilepsy
KD Ketogenic diet
LCT Long chain triglyceride
LGIT Low glycaemic index treatment
MAD Modified Atkins diet
MCT Medium chain triglyceride
MCTKD Medium chain triglyceride ketogenic diet
Modified KD’s All modified KD’s, i.e. MAD, LGIT and MKD (umbrella term)
MKD Modified ketogenic diet

Collaborator
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1.0 What is a KD?

Figure 1 represents the relative proportions of energy from macronutrients typically consumed in the human diet. Intake of a high proportion of carbohydrate in comparison to fat and protein combined results in the production and utilisation of glucose as the major source of fuel for the body when sufficient food is eaten.

In comparison, macronutrient distribution and content is manipulated in the KD so energy from fat significantly predominates over that from carbohydrate and protein combined (Figure 2).

The very high fat content of the KD produces ketones from dietary fatty acids which are used as the main energy source instead of glucose from carbohydrates i.e. the KD is designed to be ‘keto-genic’ – ketone generating. Provided carbohydrate is restricted, adequate protein included and sufficient fat consumed to meet individual energy requirements, glucose release from muscle and organ breakdown is minimised, in favour of ketone production.

In the liver, beta oxidation of fatty acids produces acetyl-CoA for conversion to ketones - ketogenesis. In the liver, ketones are transported to organs and tissues via the bloodstream and absorbed into cells. Here they are converted back into acetyl-CoA and then enter the Kreb’s cycle for oxidation in the mitochondria to energy.

History and development of the KD

The response of the body to fasting and starvation is a ‘metabolic shift’ from the production and usage of glucose predominantly from dietary carbohydrates to ketones generated from fat stored in adipose tissue (Figure 3).

Following observations dating back to the 5th century BC by Hippocrates that fasting led to seizure reduction, the KD was designed in the 1920’s to replicate this metabolic process. The KD proved efficacious in the management of epilepsy in children and adults. It was widely used prior to the increased availability of anti-epileptic drugs (AEDs) in the late 1930’s. Then, seen as complex in comparison, its use declined until the early 1990’s when interest in the application of the KD began to re-emerge as an adjunct or alternative to medication.

N.B. the degree of ketosis achieved via the KD is an individual response to an individual dietary regime. It does not always correlate with successful seizure control or other improvements in well being – both high and low ketone levels can produce equally good results.
2.1 Use of the KD

The KD has proven efficacy in the dietary management of:

- **Drug resistant epilepsy (DRE):** At least one-third of all patients with epilepsy fail to respond adequately to AEDs, and in addition to lack of seizure reduction and control, side effects from medication can contribute towards a poor quality of life9, 10. Studies on the use of the KD in children and adults with DRE show the proportion achieving 50% or greater reduction in seizure frequency ranges from 35– 56%, and that 0-15% attain seizure freedom11.

The exact mechanism (or mechanisms) by which the KD can be successful in the dietary management of DRE has yet to be fully elucidated. However, the consumption of a high proportion of daily energy requirements from fat combined with a low intake of carbohydrate appears crucial. It is likely the benefits of the KD, which in addition to control of epileptic symptoms may include, for example, improvements in cognition, sleep, developmental progress and well-being, are due to more than ketosis alone1, 2, 7, 8.

- **Inherited neurometabolic disease e.g. Glut-1 DS:** A genetic defect in primary glucose transport via the Glut-1 transporter enzyme across the blood brain barrier reduces its availability as a fuel source within the brain, resulting in seizures, complex movement disorders and global developmental delay. A range of phenotypes from mild to severe are exhibited and the condition is diagnosed from infancy through to adulthood12.

Glut-1 DS was first described in 1991 by De Vivo and colleagues13. This coincided with renewed interest in the KD and its application. The KD is now central to the effective management of Glut-1 DS as the ketones generated are able to enter the brain directly to provide an alternative and efficient energy supply. This promotes brain growth and development in infancy and early childhood, and facilitates more normal neuronal function and symptom control14.

The KD now and in the future

Renewed interest and use of the KD, together with associated clinical and scientific research, has been gathering momentum up to the present day and looks set to continue into the future. It has further established the efficacy of the KD in the dietary management of drug resistant epilepsy and neurometabolic diseases, and global guidelines and resources for its implementation for these conditions are now widely available1. Use of the KD may also have potential application in cancer and other neurological conditions15 - 18.

There are 5 versions of the KD in clinical use worldwide1, 19

The classical KD (CKD) and medium chain triglyceride KD (MCTKD) are the traditional, strict versions of the KD. ‘Modified’ versions of the KD are also available- the modified Atkins diet (MAD); Low Glycaemic Index Treatment (LGIT) and modified KD (MKD) - which were developed to be less prescriptive, more flexible and accessible approaches for implementation of the original KD. Refer to section 3.0. for details of each variant.

Although there are slight differences in the proportions of macronutrients and how each version is calculated and applied, all are very high in fat, low in carbohydrate, provide adequate protein and are designed to achieve ketosis.

Choice and application of the versions of the KD varies by country, keto-team, clinician or dietitian, and is usually based on factors such as history, experience and familiarity of use.

The use and application of the KD is evolving to incorporate the different approaches, features and attributes of each of the versions to optimally meet individual requirements and lifestyle.

An overview of a typical journey for a patient (non-infant) on the KD is shown in Figure 4.
2.2 Foods used in the KD

FAT provides fatty acids for conversion to ketones and should meet the majority of daily energy requirements. Sources are oils, butter, ghee, maroquin, mayonnaise, full-fat cream cheese, double (heavy) cream and MCT oil and emulsion.

PROTEIN, ideally from high biological value (HBV) sources, (meat, poultry, fish, eggs and dairy food) is essential for growth and tissue repair. Plant sources of protein, for example, soya, beans, seeds and nuts can be used provided an adequate amino acid intake is achieved and carbohydrate contribution accounted for.

CARBOHYDRATE mainly from small portions of vegetables and salad, berry fruits, nuts and seeds. These foods also provide FIBRE.

Foods very high in carbohydrate are avoided in a KD

- Foods high in natural or added sugars e.g. confectionary (candies) and chocolate, dried fruit, fruit juices, sweetened, carbonated drinks and cordials containing sugars, jams, honey, ice cream.
- Foods high in starch with or without natural or added sugars e.g. potato chips (french fries) and crisps, cakes, biscuits, puddings, pies and pastry products.

Other foods that can be incorporated into the KD in measured amounts

Foods, such as bread, milk, unsweetened plain/natural or Greek yogurt, pasta and rice, fruit and vegetables can be incorporated into the KD by calculation and weighing or by the use of food choices (exchange) system. This means that in addition to extending the range and choice of foods, favourite or familiar items can be included as part of meals, snacks and as ingredients in recipes. Although portion sizes will be much smaller than typically eaten in a normal diet, even tiny quantities can aid acceptability, palatability and long term compliance with a KD.

N.B. The carbohydrate contribution may be counted in some versions of the KD.

Food items usually included in the KD

- Water and sugar-free fluids e.g. carbonated drinks and cordials (not containing fruit juice), herbal and black teas, coffee.
- Unsweetened plant milks, e.g. soya, coconut, almond.
- Salt, pepper, carbohydrate-free flavourings, stocks and essences.
- Permitted, carbohydrate-free sweeteners - liquids, powders or tablets.
- Fresh and dried herbs and spices. N.B. The carbohydrate contribution may be counted in some versions of the KD.
## 3.0 Comparison of the 5 versions of the KD, and considerations

### Traditional

**History**
The original KD from the 1920’s and 30’s prior to greater availability of AEDs. Use then declined, but revived during the 1990’s.
Extensively and successfully used worldwide, especially in young children.

**Key points**
A ratio expresses the proportion of fat to protein plus carbohydrate combined, by weight in grams.
E.g. a 4 to 1 CKD indicates for every 4g of fat there is 1g of protein and carbohydrate combined.
Although the higher ratios of 4 to 1 and 3 to 1 are potentially the most ketogenic, a lower ratio of 2 to 1 may be as efficacious for seizure reduction.

Once individual daily energy and protein requirements (which form the basis of the dietary prescription and calculation) are determined, the target ratio is chosen.
The amounts of fat, protein and carbohydrate in grams required are calculated, then divided up evenly into meals and snacks so the ketogenic ratio is the same over the day.

**Version of the KD (and key references for description and implementation)**

<table>
<thead>
<tr>
<th>Version</th>
<th>Description and Implementation</th>
</tr>
</thead>
<tbody>
<tr>
<td>CKD1, 20, 32</td>
<td>The original KD from the 1920’s and 30’s prior to greater availability of AEDs. Use then declined, but revived during the 1990’s. Extensively and successfully used worldwide, especially in young children.</td>
</tr>
<tr>
<td>MCTKD33, 34, 21</td>
<td>Developed by Huttenlocher et al in the 1970’s as a modification of the CKD, using medium chain triglyceride (MCT) oil to replace a proportion of long chain triglycerides (LCT) and take advantage of the greater ketogenic potential of the medium chain fatty acids (MCFA) C8 (octanoic) and C10 (decanoic).</td>
</tr>
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</table>

### Modified

**History**
Developed from observations that:
- Blood glucose levels are stable on the CKD and during fasting.
- Diets with a low glycaemic index (GI) are linked with favourable health measures and outcomes.
- Excessive intakes of high GI foods may provoke seizures in some people with epilepsy.

**Key points**
Only carbohydrate foods (40 – 60g per day including fibre), with a GI of ≤ 50 are permitted.
The level of carbohydrate in the LGIT has been chosen to reflect that 50g per day is the physiological maximum below which the body will use fat as its primary energy source.
Energy requirements are typically assessed before the diet is started (especially for children), and advice given on protein and fat intakes to ensure consumption of appropriate amounts.

**Version of the KD (and key references for description and implementation)**

<table>
<thead>
<tr>
<th>Version</th>
<th>Description and Implementation</th>
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</thead>
</table>
| LGIT1, 28 | Developed from observations that:
  - Blood glucose levels are stable on the CKD and during fasting.
  - Diets with a low glycaemic index (GI) are linked with favourable health measures and outcomes.
  - Excessive intakes of high GI foods may provoke seizures in some people with epilepsy. |
| MAD35, 36 | Developed as a result of seizure reduction observed by patients with epilepsy using the low carbohydrate, high protein Atkins diet originally intended for weight reduction. |
| MKD37 | Has evolved as a ‘hybrid’ of features from the other 4 versions of the KD, e.g. incorporating MCT, using low GI carbohydrate, and offering a more flexible approach to food selection. |

### Ratio by weight of fat to carbohydrate and protein combined

The typical human diet is approximately 0.3 to 1

<table>
<thead>
<tr>
<th>Ratio</th>
<th>Description</th>
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<tbody>
<tr>
<td>e.g. 2 to 1, 2.5 to 1, 3 to 1, 4 to 1</td>
<td>Approximately 1.6 to 1</td>
</tr>
<tr>
<td>Approximately 1.6 to 1</td>
<td>Approximately 1 to 1</td>
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</table>

### Comparisons of approximate ratio of macronutrients by energy intake from fat, protein and carbohydrate

- Diagrams showing the approximate ratio of macronutrients by energy intake from fat, protein, and carbohydrate.
### Comparison of the 5 versions of the KD, and considerations, continued

#### Macro and micronutrients

<table>
<thead>
<tr>
<th>Fat - LCT</th>
<th>Fat - MCT</th>
<th>Essential fatty acids (EFA) - omegas 3 and 6</th>
<th>Protein</th>
<th>Carbohydrate</th>
<th>Micronutrients</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Proportionally, intakes of saturates, mono or polyunsaturates will predominate depending on the particular high fat foods most frequently consumed, e.g. saturates from butter and double (heavy) cream, monounsaturates from olive oil use or polyunsaturates from seed oils, e.g. sunflower, safflower or rapeseed (canola).</td>
<td>• Pure coconut oil consists predominantly of C12. Although it can be classed as a MCF A, C8 and C10 are considered to have the greater ketogenic and anti-epileptic potential.</td>
<td>• Due to the fats typically consumed in the KD, intakes of omega 3 fatty acids can be below that recommended for health. Use of different fat sources provides balance, together with inclusion of vegetable oils (e.g. walnut, flaxseed, linseed) and foods (oily fish, fortified eggs, seeds, nuts and green vegetables) naturally high in omega-3 fatty acids.</td>
<td>• For all versions of the KD sufficient protein and essential amino acids must be included for growth, function, repair and replacement of body tissues. To ensure adequate intakes: • Include HBV protein (meat, fish, eggs, cheese) in each meal every day. • Avoid sole or over-reliance on plant sources of protein (vegetables, tubers, grains, soya, pulses, peas, beans, lentils or nuts) to meet requirements.</td>
<td>• Daily carbohydrate intake is influenced by protein and energy needs and diet ratio. E.g. on a higher protein, lower energy, 3 to 1 or 4 to 1 ratio KD there is less carbohydrate compared to a lower protein, higher energy 2 to 1 ratio. This may influence acceptability and compliance as it affects palatability.</td>
<td>Micronutrient content is inherently low and/or inadequate in all versions of the KD. A comprehensive, daily supplement is to meet daily requirements is advised, or as per keto-team policy.</td>
</tr>
<tr>
<td>• Prudent inclusion of a variety of sources of LCT from both animals and plants, provides a balanced mixture of all three types of fatty acids in the context of cardiovascular and metabolic health.</td>
<td>• MCT can cause adverse gastrointestinal symptoms, needs to be introduced slowly and carefully. Further information on the use of MCT in the KD and establishing gastrointestinal tolerance is available for downloading from <a href="http://www.nestlehealthscience.com/Vitaflo/VIA">www.nestlehealthscience.com/Vitaflo/VIA</a>.</td>
<td>• EF A’s are not present in MCT and intakes can be low. Although some will be provided via LCT, inclusion of oil and food sources of omega 3 is advised, as per the CKD.</td>
<td>• A free choice of protein food(s) can be made but moderate intakes are advised, i.e., normal sized portions. If eaten to excess, protein foods, due to their high palatability, may replace those providing fat and compromise ketosis.</td>
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<td></td>
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<td>• C8 and C10 are predominant in the MCT oils and emulsions available for clinical use and are therefore recommended in preference to coconut oil as a source of MCT and MCF A in the KD.</td>
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<td></td>
<td></td>
<td>• N.B. The MCF A CS and C10 are saturated. However, evidence suggests they have potentially beneficial health effects in comparison to saturated fatty acids with longer chains.</td>
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<tr>
<td></td>
<td></td>
<td>• MCT can be introduced and if taken at regular intervals with food (as part of daily meals and snacks) may help ‘boost’ ketone production without needing to increase the ketogenic ratio.</td>
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<tr>
<td></td>
<td></td>
<td>• The amount of MCT calculated for the patient is distributed evenly amongst meals, snacks and at bedtime, i.e. consumed regularly throughout the day, and always with food.</td>
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<tr>
<td></td>
<td></td>
<td>• Once established on diet, MCT can be introduced and if taken at regular intervals with food (as part of daily meals and snacks) may help ‘boost’ ketosis without the need for an increase in LCT or reduction in carbohydrate intake.</td>
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<td>• N.B. The MCF A C8 and C10 are saturated. However, evidence suggests they have potentially beneficial health effects in comparison to saturated fatty acids with longer chains.</td>
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4.6 Pre-KD assessments and clinical outcomes

The KD is complex and can have side effects. Before a patient is started on a KD, undertake the following:

- Clinical, anthropometric and dietary assessment (4.1 - 4.3).
- Pre-KD clinical and/or dietary intervention, if required (4.1).
- Measurement of clinical outcomes to provide baseline data on the patients current health status. These can then be used as serial assessments to determine change or benefit, e.g. after 3, 6 and 12 months. Specific KD outcome measures may include seizure frequency and severity, dietary adherence, adverse effects, anthropometry, quality of life.
- Obtain commitment from the patient and/or parents/caregivers to follow the KD for the recommended trial period of 3 months to determine its efficacy.
- KD training for the patient and/or all involved in their care, e.g. family, teachers, respite care staff.

4.1 Clinical and dietetic assessments

**Does your patient currently have this?**

**Relevance to the KD**

**Initiate BEFORE starting a KD**

<table>
<thead>
<tr>
<th>Gastrointestinal disorder(s)</th>
<th>Constipation</th>
</tr>
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<tbody>
<tr>
<td>Gastro-oesophageal reflux (GOR), delayed gastric emptying, oesophageal and foregut dysmotility.</td>
<td>Constipation is linked to low dietary fibre and fluid intakes. Carbohydrate restriction limits consumption of fibre-containing plant foods such as fruits, vegetables and cereals. In addition, being constipated may be associated with seizures.</td>
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<tr>
<td>Prophylactic laxatives (carbohydrate free) before starting and whilst on the KD may be indicated to prevent and manage constipation.</td>
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<table>
<thead>
<tr>
<th>Physical feeding problems</th>
<th>Inadequate fluid intake due to poor/unsafe swallow</th>
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<tbody>
<tr>
<td>Oro-motor impairment causing:</td>
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<tr>
<td>• Difficulties with chewing, biting &amp; swallowing.</td>
<td>Low fluid intake on the KD is linked to constipation and urolithiasis. Foods high in fat are low in moisture. Those that typically contribute water in a normal diet, e.g. vegetables and fruits, are restricted. Thus, the inherent fluid content of a KD is low.</td>
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<tr>
<td>• Increased aspiration risk.</td>
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<td>• Prolonged feeding times.</td>
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<tr>
<td>The KD has a soft, semi solid texture due to its high fat content. This consistency can be well suited to those with feeding difficulties. A daily pattern of small, frequent meals and snacks, often ideal for those with feeding issues, can also help to achieve and maintain ketosis on the KD.</td>
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<td>The high energy density of fat means that relative to a normal diet, ketogenic meals and snacks are much smaller in size. This reduction in volume may help those who struggle to consume adequate food by facilitating quicker and more efficient feeding and an improved nutritional intake.</td>
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<tr>
<td>The restricted range of foods permissible on the KD may suit fussy eaters. Personal and family food preferences can be incorporated into the KD and may aid compliance and acceptance.</td>
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<tr>
<td><strong>Mandatory assessment by specialist feeding and/or gastroenterology team.</strong></td>
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<tr>
<td><strong>Action BEFORE starting KD</strong></td>
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<tr>
<td><strong>Implementation strategies and interventions before starting a KD.</strong></td>
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<td><strong>Ensure they are successful prior to starting the KD.</strong></td>
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<table>
<thead>
<tr>
<th>Food allergies e.g. milk, egg.</th>
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<tbody>
<tr>
<td>Food intolerance e.g. lactose, wheat.</td>
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<tr>
<td>Coeliac disease</td>
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<tr>
<td><strong>Ensure correct diagnosis made and food exclusion actually required.</strong></td>
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<table>
<thead>
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<td></td>
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<tr>
<td><strong>Fat is inherently low in micronutrients; combined with restrictive food choices, the risk of nutritional deficiencies is increased.</strong></td>
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<tr>
<td><strong>Specific micronutrient and/or comprehensive supplementation should be started before the KD commences. Note: Daily micronutrient supplementation is recommended for all patients on the KD.</strong></td>
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<tr>
<th><strong>Poor nutritional quality of current diet.</strong></th>
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<tr>
<td><strong>At risk of/know micronutrient deficiency.</strong></td>
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**N.B. If any of these issues continue or develop once on the KD, refer for specialist assessment and advice.**

**Expectations**

- Patients and/or parents/caregivers need to be motivated and have decided they definitely want to embark on a KD.
- The KD doesn’t always work, although for some individuals improvements in seizure management and quality of life may be better than anticipated.

As part of the assessment and pre-KD preparation process, realistic goals for dietary management and clinical outcomes should be discussed and agreed between the keto-team and patient and/or parents/caregivers.
4.2 Anthropometric

<table>
<thead>
<tr>
<th>Children and adolescents</th>
<th>Adults</th>
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</thead>
<tbody>
<tr>
<td>Weight (kg)</td>
<td>Height (cm)</td>
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<tr>
<td>If aged between 1 and 2 years - head circumference (cm)</td>
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<tr>
<td>Optional - calculate Body Mass Index (BMI).</td>
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<tr>
<td>Plot on a growth chart (along with any historical growth data, if available, for reference).</td>
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<tr>
<td>N.B. Children’s growth velocity and expected weight gain may slow on the CKD, especially over time1, 49.</td>
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</table>

Some initial weight loss may be beneficial for initiation and maintenance of ketosis at the start of a KD. In children, aim for stabilisation of weight over time to allow for growth.

Advise on increasing energy intake by adding extra fat into the current diet - both to promote weight gain and to get used to eating more before formally starting the KD.

- Once on the KD, regular anthropometric and dietetic assessment to monitor clinical outcomes is especially important for children and those with pre-existing compromised nutritional status, feeding problems and/or impaired mobility and activity levels, to help ensure dietary intake (particularly of protein) is appropriate.

- If the KD proves effective, seizure reduction may lead to an increase or decrease in energy requirements. Likewise, changes in mobility and activity, e.g. improvements in ambulatory skills, may alter individual needs and impact nutritional status over time.

**Overweight pre-KD**

Some initial weight loss may be beneficial for initiation and maintenance of ketosis at the start of a KD. In children, aim for stabilisation of weight over time to allow for growth.

**Underweight pre-KD**

Advise on increasing energy intake by adding extra fat into the current diet - both to promote weight gain and to get used to eating more before formally starting the KD.

4.3 Dietary assessment

- For the CKD and MCTKD, detailed and specific information about current nutritional status and usual diet enables estimation of actual daily energy needs (which may differ from those recommended for age) and calculation of quantities of macronutrients for meals and snacks. This information facilitates practical guidance to be given by the keto-team and can help achieve greater success in diet implementation and efficacy1.

- In comparison, for the modified versions of the KD (MAD, LGIT or MKD), pre-diet evaluation may be less formal.

- Before starting the MAD, no diet history is taken from the patient nor are they advised to have a specific daily energy intake34, 35. This is to guide the advice given to the patient on appropriate daily macronutrient intakes (especially fat) to meet these needs, and to promote ketosis.

Examples of dietary assessment tools:

- 3-4 day diet diary filled in by the patient and/or carer
- Verbal diet history (previous 24 hours and/or longer retrospective period) and/or
- Food frequency questionnaire

Dietary assessment identifies:

- Specific details of foods and drinks consumed i.e. those avoided and restricted; textures (important for those with feeding problems), preferred food combinations, flavours, presentations, portion sizes.
- Usual eating and drinking intake, habits and patterns.

- How foods and drinks are prepared, and by whom.
- Sources and availability of foods, eating venues e.g. home and institutions (school, college, respite care), takeaways, cafés, restaurants.

Action points when planning a KD:

Include familiar and favourite foods and drinks where possible to aid compliance.

If eating habits are irregular, advise that a consistent daily meal and snack pattern is established pre-KD as this will aid maintenance of ketosis.

Aim to normalise fluid intakes as:

- Too little is linked to side effects, e.g. constipation, urolithiasis.
- Or
- If excessive - especially in young children - this may decrease appetite and food intake, affecting the ability to consume the KD properly and achieve ketosis.

Educate and provide information to all involved in preparing foods, drinks and feeding the patient. This is vital prior to starting a KD.

Give information and advice on making correct food and drink choices when eating out and away from home.

Information gained from clinical, anthropometric and dietetic assessments, and from the patient and/or parents/caregivers, will aid choice of the most suitable version of the KD for them (4.4)1.
### 4.0 Assessment and clinical outcomes, continued

#### 4.4 Choosing a version of the KD for your patient

<table>
<thead>
<tr>
<th><strong>Traditional Versions</strong></th>
<th><strong>Suggestion</strong></th>
<th><strong>To consider</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Is a younger child.</td>
<td>CKD</td>
<td>Labour intensive - all foods need weighing and measuring and all meals and snacks need calculating. Lots of fat, very little carbohydrate and protein; meals look small.</td>
</tr>
<tr>
<td>Would prefer a structured format and detailed, precise instructions to follow.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Has a poor appetite, can be a fussy or slow eater or self restricts; prefers small meals.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Needs an enteral feed taken orally or via a feeding tube.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Is of any age (children, adolescent and adults).</td>
<td>MCTKD</td>
<td>Labour intensive - all foods need weighing and measuring and all meals and snacks need calculating. MCT is introduced gradually, building up to the required daily quantity. Hence, it may take a while for the MCTKD to be fully tolerated and established before efficacy is determined. Must be prepared to take MCT regularly throughout the day, with food.</td>
</tr>
<tr>
<td>Would prefer a structured format and detailed, precise instructions to follow.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Likes carbohydrate and protein foods.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Needs more protein than achieved from a CKD.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Is willing to use a food choices (exchange) system for fat, carbohydrate and protein.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Needs an enteral feed taken orally or via a feeding tube.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

#### Modified Versions

<table>
<thead>
<tr>
<th><strong>Modified Versions</strong></th>
<th><strong>Suggestion</strong></th>
<th><strong>To consider</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Is an older child, adolescent or adult. Needs a less restrictive diet than a CKD or MCTKD i.e. less weighing of foods and more flexibility with food choices and quantities.</td>
<td>LGIT</td>
<td>40 - 60g carbohydrate (including fibre) each day from foods with a GI &lt; 50.</td>
</tr>
<tr>
<td>They and/or their parents/caregivers can cope with a less structured KD.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Eats out or is away from home regularly.</td>
<td>MAD</td>
<td>Very restrictive carbohydrate intake at first.</td>
</tr>
<tr>
<td>Wants 'normal sized' and 'normal looking' meals.</td>
<td>MKD</td>
<td>Has the potential to be the most 'tailored' to the patient's nutritional and lifestyle requirements by incorporating elements of the other 4 versions of the KD.</td>
</tr>
<tr>
<td>Willing to use a food choices (exchange) system for fat and carbohydrate to assist with meal planning and portion sizes from the outset or as an aid to compliance.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Traditional Versions**

**Suggestion**

- To consider

**Modified Versions**

**Suggestion**

- To consider

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**For the KD to be successfully implemented and followed, patients and/or parents/caregivers need the ability and confidence to:**

1. Measure foods and portion sizes accurately and consistently.
   - For the CKD and MCTKD, by using digital electronic scales to weigh foods in grams.
   - For the modified versions of the KD (LGIT, MAD and MKD), using household measures (i.e. spoons, cups) and/or digital electronic scales to weigh foods in grams, as directed by the keto-team.
2. Choose, shop for, cook and prepare meals, snacks and recipes from foods as calculated and advised by the keto-team.
3. Use information on food labels to help guide the selection and inclusion of foods for meals and snacks, e.g. from:
   - The ingredients list to identify sources of macronutrients, especially carbohydrate and fat.
   - The nutrition information box, to identify amounts of macronutrient per 100g of food, so that specific quantities in grams can be calculated, either as portions or as part of the food choices system.
4. Follow and implement written dietary advice and instructions provided by the keto-team.
5. For those on enteral feeds, prepare a KD feed recipe as calculated by the keto-team.

**Education and resources for the KD can be provided via:**

- Face-to-face meetings and teaching sessions, on an individual, family or group basis.
- The telephone, live video conference calls, emails or texts.
- Printed copies of the individual meal plan, meal and snack suggestions and recipes specifically prepared for the patient.
- Opportunity to prepare and try out recipes, meals and snacks beforehand, at home or KD cookery events run by the keto-team.
- Information booklets, leaflets, recipes and advice sheets written by the keto-team and/or from charities and organisations providing advice and support for the KD when used for epilepsy and neurometabolic disorders, e.g. Matthew’s Friends, www.matthewsfriends.org, or The Charlie Foundation, www.charliefoundation.org
- The Glut-1 Deficiency Foundation, www.glidfoundation.org
- Links to patient-friendly ketogenic diet websites, e.g. www.myketogenicdiet.co.uk
- ‘Buddy-ing up’ with others who have used, or are using, the KD who are willing to help those new to the diet, by meeting in person, via social media or on the telephone.
- Provide KD information and training to everyone involved with and caring for the patient, e.g. relatives and friends, education and respite care staff.
- Encourage patients and/or caregivers to practice making up meals, snacks and recipes to sample before the KD formally starts, to build confidence in food choice and preparation. A period of preparation beforehand is recommended to promote acceptance of and adherence to the KD - so represents time well spent!
6.0 Making dietary changes before starting the KD

Changing over to a diet very high in fat and containing minimal carbohydrate is demanding, and requires many lifestyle adjustments by patients and parents/caregivers. In addition to restricted food choices, requisite changes to macronutrient proportions in meals and snacks alters their taste, texture and size. This can compromise tolerance and acceptance of the KD and make implementation more challenging.

To aid this transition, simple alterations can be made to the patients usual diet for a short period, e.g. 1-2 weeks, prior to them starting the KD. Information from the dietetic assessment (Section 4) can help tailor individual advice to avoid compromising energy and nutritional intakes.

Examples of pre-KD dietary changes are:

- Omit foods and drinks high in sugar, e.g. sweets, chocolate, cakes, biscuits, puddings, icecream.
  - Swap high sugar cordials, fizzy pop and fruit juice to sugar-free varieties or plain water and
  - Use sweeteners instead of adding sugar or honey to foods and drinks, or low sugar or sugar free alternatives to sugary yogurts, desserts and breakfast cereals.

- Reduce intakes of foods high in starch, e.g. potatoes, rice, pasta, bread, cereals, beans and pulses.
  - Give practical advice on portion sizes and/or suggest full or partial replacement with low carbohydrate vegetables and salads.
  - Milk (whole, semi-skimmed or formula) is generally avoided in the KD, although small amounts can be included in the MCTKD. Exchange milks for low carbohydrate, unsweetened soya or nut milks or diluted double (heavy) cream.

- Increase fat intake, (ideally from a variety of animal and plant sources, to provide a mixture of saturated, monounsaturated and poly-unsaturated fatty acids) by:
  - Frying foods.
  - Using full fat dairy products such as Greek yogurt and cheese (both useful as snacks to replace sugary foods).
  - Adding extra fat to foods, e.g. butter, margarine, cream cheese or oil to potatoes, vegetables and bread; mayonnaise or oil based dressing to salads, and cream (double or heavy) to fruit, low sugar desserts or yogurt.

In addition, dietary habits that should be established prior to starting a KD are:

- Eating meals and snacks regularly during the day. This can help promote a steady and sustained ketosis when on the KD.

- Achieving a daily fluid intake that meets individual requirements for hydration. This is important once on the KD, due to its inherent low fluid content.

- Taking a daily micronutrient supplement. This is to ensure that despite the restrictive nature of the KD, nutritional requirements will be adequately met.

References


References, continued


