



Ketogenic Dietary Therapies
Information • Training • Research • Support
Registered Charity No. 1108016

AN INTRODUCTION TO
Medical Ketogenic
Dietary Therapies

The Matthew's Friends Organisation

The Background:

Matthew Williams suffers with a catastrophic form of epilepsy called 'Dravet Syndrome' and his seizures started when he was 9 months old. Emma, his mother, asked if Matthew could try the ketogenic diet when he was 2 years old, but she was told the diet didn't work. So she battled on, trying innumerable medications which didn't help Matthew's seizures and which caused devastating side effects. Six years later, Professor Helen Cross began a research trial of the ketogenic diet at Great Ormond Street Hospital (GOSH). Matthew and 144 other children with severe epilepsy were enrolled on the trial.

Within 2 weeks of starting the diet Matthew's seizures had reduced by 90% and within 8 months he was off all medication. Sadly for Matthew, the damage had been done. Years of seizures had caused terrible brain damage, his family had broken apart and Emma was now a single mum to Matthew and his younger sister Alice. Inspired by Matthew, she set up the Matthew's Friends charity in 2004 to provide information, education, training, resources and support for other families.



The charity, which was started at Emma's kitchen table, grew at a phenomenal rate and in 2011 Emma realised her dream of opening the Matthew's Friends ketogenic therapies clinic employing an expert medical team to provide the therapy and train other clinical teams, as well as looking into new research possibilities.

Today

The charity invests considerable funds for clinical research into ketogenic therapy and its applications, as well as training and educational programmes. The Charity provides free starter packs and appropriate ketogenic samples and goods to families in the UK and Ireland and we are providing funding for numerous NHS centres around the UK. 2011 saw the registration of Matthew's Friends New Zealand and 2014 saw the registration of Matthew's Friends Canada, as well as being involved with over 20 other countries worldwide in supporting the use of ketogenic therapy.

The organisation has expanded its remit to help adults that are failing anti-epileptic medication and is also now working in the field of certain types of brain cancer and metabolic disorders such as Glut 1 Deficiency. 2016 saw the launch of the first 'Matthew's Friends KetoCollege'.

“A proven medical therapy for epilepsy based on a diet that is high in fat, adequate in protein and low in carbohydrate”.

What is a Ketogenic Dietary Therapy?

Ketogenic diet is more than its title suggests - it is NOT some fad diet that is currently 'fashionable'. It is a proven medical therapy for epilepsy based on a diet that is high in fat, adequate in protein and low in carbohydrate. It MUST be managed by a qualified ketogenic dietitian in partnership with neurologists / doctors. It is clinically proven to be highly effective in managing difficult to control epilepsies, as well as metabolic disorders such as GLUT1 deficiency, with research currently being conducted looking at the diet as a therapy for a range of long term neurological diseases and cancer.

The diet was first developed in the USA in the 1920's and was used for children and adults. With the advent of newer anti epileptic medication in the 1930's-1970's interest in the diet waned. The 1990's, however, saw a resurgence of interest in the diet for children. In the 1970's the MCT (medium chain triglyceride oil) version of the diet was also introduced.



Types of Ketogenic Therapy

All types of the diet are based on regular fresh food ingredients; meats, fish, eggs, nuts, seeds, cheese, vegetable oils, butter, cream, vegetables and fruits.

- Classical ketogenic diet - where carbohydrate, fat and protein amounts are all measured and carefully distributed to maintain a similar balance (ketogenic ratio) at meals and snacks.
- MCT ketogenic diet - where carbohydrate, fat and protein amounts are all measured and medium chain triglyceride oil e.g. refined coconut oil is included with each meal / snack. This enhances ketosis and allows more carbohydrates in meals and snacks.
- Modified ketogenic diet and Modified Atkins diet - not so restrictive on protein foods but requires measurement of carbohydrate foods and adequate portions of fats at meals.
- Low Glycaemic Index Treatment – very similar to the Modified ketogenic diet approach in terms of measuring carbohydrate and encouraging fats but restricts the carbohydrate sources to those with a glycaemic index of 50 or below.

Ketogenic dietary therapies can be administered as a normal oral diet, via a bottle feed, or tube fed and specific formulas are available.

These diets can be tailored to patients specific requirements which could mean incorporating elements of all the above diets as we seek to optimise the effects.

"The "Holy Grail" of the ketogenic diet is for a patient to be initiated on the diet, become seizure free, reduce/remove the amount of anti-epileptic medication taken, wean the diet off after a period of 2 years and STAY seizure free."

"If it means that you have less or no seizures and a better quality of life with more choice and freedom – it is certainly worth a trial of 3 months".

Ketogenic Therapy for Epilepsy - How the Diet works

There is much research in this area! The diet appears to "mimic starvation" by using fat as an alternative fuel source for the body, producing ketones. These ketones and the associated biochemical changes in the brain, can have an anti-convulsive effect.

The "Holy Grail" of the ketogenic diet is for a patient to be initiated on the diet, become seizure free, reduce/remove the amount of anti-epileptic medication taken, wean the diet off after a period of 2 years and STAY seizure free. This DOES happen for some, but there are also other degrees of success on the diet:

- Reduction in number of and intensity of seizures
- Reduction in drugs and their subsequent side effects
- Increased alertness
- Improvement in behavioural problems
- Improvement in learning ability
- QUALITY OF LIFE!



Fears and Misconceptions

Arguments against using the diet usually consist of the following:

"The diet is difficult"

It depends what you mean by "difficult" – it's difficult to watch your child seize. It's difficult to watch your child in a drugged up haze. It's difficult to feel that you've "lost" the child you once had to epilepsy. Compared to these, spending time in the kitchen, actively involved in your child's care is a piece of cake! We don't want to belittle the amount of effort needed to calculate or administer the diet – Initially you can feel like you're never out of the kitchen and it can be a daunting prospect but we just want to put this effort in context.

Also for adults whose daily lives are severely impaired by poorly controlled epilepsy and the side effects of many medications; finding that it affects their ability to work, socialise, drive and just live from day to day. Is it really more difficult to change some of the things you eat? If it means that you have fewer seizures or no seizures and a better quality of life with more choice and freedom - it is certainly worth a trial of 3 months.

“It is unpalatable”

ALL forms of these dietary therapies are high fat, adequate protein and low in carbohydrate and in the early days of the diet the above was true – cups of oil had to be drunk and spoonfuls of butter needed to be eaten – that is NOT the case anymore. As much as possible we work from the usual meals made at home although in some instances, we do have to change some of the ingredients to make the meal more ‘ketogenic’.

There are many good ketogenic recipes available now by way of recipe books for mainstream lower carbohydrate diets and specialist ketogenic diet websites. At Matthew’s Friends website (www.matthewsfriends.org) we have an extensive recipe section, with Handy Guides and tips as well as our own YouTube Cooking channel, giving tutorials and guidance on recipes that are easy to make and delicious to eat.

“It Won’t Work for Everyone”

That is true – sadly the diet doesn’t work for everyone, but it does have a good success rate for drug resistant epilepsies, as good as any new anti-epileptic drug that is currently on the market and remember, the drugs don’t work for everyone, VNS won’t work for everyone and surgery is not always an option for a patient. We can usually tell within 3 months whether a ketogenic dietary therapy is going to be helpful or not.



“The positive effects may not last”

You may be told that “only rarely do the effects last more than 12 months” – many can testify that this is often not the case, and if the usual level of control does start to deteriorate, expert fine tuning will hopefully bring things back on track.

“The diet won’t work for adults”

There is published evidence stretching back to the 1930’s and recent international experience showing that diet manipulation can have an impact on seizures and associated symptoms in adults. It is suggested that:

Approximately 40-50% of adults on supervised ketogenic diet therapy will achieve a 50% reduction in seizures; some benefiting more or less than this.

Other less quantifiable benefits commonly reported are:

- An improvement in energy levels and general ‘zest for life’
- A shorter hangover time after seizures
- An improved clarity of thought and ability to make decisions
- A feeling of gaining some control over life that, in time, leads to improved mental health

Please note that any type of ketogenic dietary therapy should only be undertaken with strict medical supervision by an experienced team.

“This treatment has been shown to be successful in treating a wide range of seizure types and syndromes although may be particularly beneficial in myoclonic epilepsies, infantile spasms and tuberous sclerosis complex”.

“A ketogenic diet can be administered to enterally-fed individuals via naso-gastric tube or gastrostomy”.

Who is ketogenic therapy suitable for?

Ketogenic therapy should be considered as a treatment for epilepsy after two appropriate anticonvulsant medications have failed to be effective or produced unacceptable side effects. This treatment has been shown to be successful in treating a wide range of seizure types and syndromes although may be particularly beneficial in myoclonic epilepsies, infantile spasms and tuberous sclerosis complex. Guidelines from the UK National Institute for Health and Care Excellence (NICE), updated in 2012, recommend ‘Refer children and young people with epilepsy whose seizures have not responded to appropriate anti-epileptic drugs to a tertiary paediatric epilepsy specialist for consideration of the use of a ketogenic diet.’ (<http://www.nice.org.uk>).

The ketogenic diet is a first-line treatment for the neurometabolic diseases glucose transporter type 1 (GLUT1) deficiency syndrome and pyruvate dehydrogenase (PDH) deficiency.



Contraindications

Ketogenic therapies are not suitable for those individuals who have the following conditions:

- fatty acid oxidation defects
- pyruvate carboxylase deficiency
- primary carnitine deficiencies,
- organic acidurias,
- defects in ketone metabolism,
- disorders that require high dietary carbohydrates as part of their treatment
- severe liver disease
- hypoglycaemia under investigation.

They should be used with caution in those with a history of:

- renal stones
- hyperlipidemia
- severe gastro-oesophageal reflux
- diabetes mellitus

Certain family circumstances may also hinder the management of the diet and also some treatments such as steroid use may also interfere with this therapy.

Although possible, ketogenic therapy will be more difficult to implement if there are pre-existing dietary restrictions and food allergies, especially if associated with behavioural feeding issues. A ketogenic diet can be administered to enterally-fed individuals via naso-gastric tube or gastrostomy.

All types of diet have been shown to be successful in the treatment of epilepsy. Choice will depend on the individual's age, dietary preference and lifestyle, taking into account palatability and ease of use for both the child/adult and the family.

The whole process becomes more familiar and easier over time, in the early weeks, it can seem like there is little time to do anything else other than simply manage the therapy”.

Preparation and initiation: Commitment to the initial three months

Ketogenic therapy is an experimental journey with the individual treatment path evolving over months. The first few weeks are generally the most challenging in that you are learning to cope with significant changes in meal planning, shopping for new foods, measuring food portions and altering cooking methods. In addition, there is the responsibility for monitoring and recording seizure and symptom changes and frequent feedback that your team will require of you.

The whole process becomes more familiar and easier over time, in the early weeks, it can seem like there is little time to do anything else other than simply manage the therapy. For this reason, it is important that the trial occurs when home life is relatively stable and you are able to give it your best attention. Your medical team will ensure that medication adjustments are managed before starting so that the only variable to change during the trial period is the food choice.

Thorough preparation, careful initiation, meticulous monitoring and close working with your team is essential to give this therapy its best chance to work.



Preparation of food

(4- 8 weeks before starting
a prescribed regime)

Once the pre-ketogenic neurological and biochemical screening has been carried out and a possible time frame for starting ketogenic therapy has been agreed, the preparations need to start. You may well be asked to attend an education session and your food records, food preferences and normal patterns of eating will be used as a basis to advise on how the ketogenic diet will be structured and what you may do in preparation for this.

“It is a good idea to look for alternative options and reduce the dependency on sweet foods BEFORE starting the ketogenic diet”

Although all diets are based around similar principles of low carbohydrate and high fat intake and all will be tailored to your individual nutritional requirements, the way in which the different protocols are implemented will vary.

Common step changes are as follows:

- Maintaining a regular meal & snack routine, becoming aware of portion control and ensuring that all food is eaten. It is also very important to evolve a strategy to manage any feeding related behavioural issues, before commencing ketogenic therapy.
- Cutting out sugar, sweets, chocolates, cakes & biscuits. It is a good idea to look for alternative options and reduce the dependency on sweet foods BEFORE starting the ketogenic diet.
- Increasing the fats and oils in meals. You may be advised to start using full fat (not low fat) products and trying out ways of increasing the amounts of fats and oils (olive oil, coconut oil, butter, mayonnaise, double cream etc) added into meals
- Trying out a few ketogenic recipes.



The diet prescription

Ketogenic dietary therapies are designed to cause a metabolic shift within the body. Fat becomes the primary fuel rather than carbohydrate, with ketone bodies replacing glucose as an energy source for the brain. Although all diets are based around similar principles of low carbohydrate and high fat intake and all will be tailored to your individual nutritional requirements, the way in which the different protocols are implemented will vary.

Dietary fine tuning

The aim of fine-tuning a diet is to establish the prescription for optimal efficacy and on-going dietary modifications are an essential component of the dietetic care. Regular home monitoring of ketone levels, weight and seizures are important. Ketone levels are a useful indication of how the body has adjusted to the diet, and you need to ensure they are not excessive. The level of ketosis associated with the best seizure control will vary between individuals.

“A ketogenic enteral feed can be used for existing tube fed patients who are to be initiated on ketogenic therapy, or for patients already established on ketogenic therapy who subsequently require tube feeding”.

Enteral feeding of ketogenic therapy

Use of an enteral feeding tube to provide all or part of a prescribed ketogenic therapy is indicated in situations where adequate oral intake is not possible due to medical conditions.

It may also be used to supplement an inadequate oral intake resulting from physical or behavioural eating problems. A ketogenic enteral feed can be used for existing tube fed patients who are to be initiated on ketogenic therapy, or for patients already established on ketogenic therapy who subsequently require tube feeding.



Side effects of ketogenic therapy

Early side effects

During initiation of ketogenic therapy there is a risk of hypoglycaemia, acidosis, dehydration and high levels of ketones. Children on carbonic anhydrase inhibitor medications (for example, topiramate or zonisamide) may have increased risk of excess ketosis and metabolic acidosis on commencing ketogenic therapy. It is therefore important to keep in close contact with your managing team in order to reduce the risks of early side effects and have any possible side effects rectified quickly and effectively.

Nutrition and growth

There is evidence of impaired growth in children on the ketogenic diet and younger children may be more at risk. Long-term follow up of children treated with the ketogenic diet in the past suggests that although growth does improve after diet treatment is discontinued, height gain can still be below expected.

Individual requirements for vitamin, mineral and trace element supplementation, together with growth will be closely monitored by your team.

“Raised blood lipids may trend back to normal with time on treatment and dietary modifications can also help to achieve healthy levels”.

Cardiovascular

Elevated blood cholesterol and triglyceride levels have been reported in children on the ketogenic diet with significant increases in atherogenic apoB-containing lipoproteins. Raised blood lipids may trend back to normal with time on treatment and dietary modifications can also help to achieve healthy levels.

Kidney stones

The reported incidence of kidney stones in children on the ketogenic diet is higher than that of the general paediatric population. Uric acid, calcium oxalate, calcium phosphate or mixed composition stones have been reported in up to 7% of children on the diet. Risk may be higher with long-term treatment and concurrent use of carbonic anhydrase inhibitors such as topiramate or zonisamide.

Gastro-intestinal

Gastro-intestinal problems are common side effects of ketogenic therapy however can usually be alleviated with dietary manipulation. Vomiting, nausea, diarrhoea, and abdominal discomfort have been reported in studies of the MCT and classical ketogenic diets. There is a risk that children with pre-existing gastro oesophageal reflux will have symptoms exacerbated by a high fat regime. Constipation is the most common reported complication of ketogenic therapy; and despite dietary changes to help lessen the problem some individuals do need additional treatment with medication.



“Raised blood lipids may trend back to normal with time on treatment and dietary modifications can also help to achieve healthy levels”.

Other side effects

Other rare side effects that have been reported with ketogenic therapy are increased infection risk, bruising, raised uric acid in the blood, bone fractures, pancreatitis, lipid-aspiration pneumonia, and cardiac abnormalities. Ketogenic diet-induced toxicity of the anticonvulsant medication sodium valproate has been reported however these treatments can be used together without problem in most cases.

Using Modified diets

The literature on side effects of ketogenic therapies has mainly studied traditional, more restricted versions of ketogenic dietary therapy. The limited reports of complications arising from use of the modified diets, suggest fewer and less serious adverse events when these more liberal diets are used.

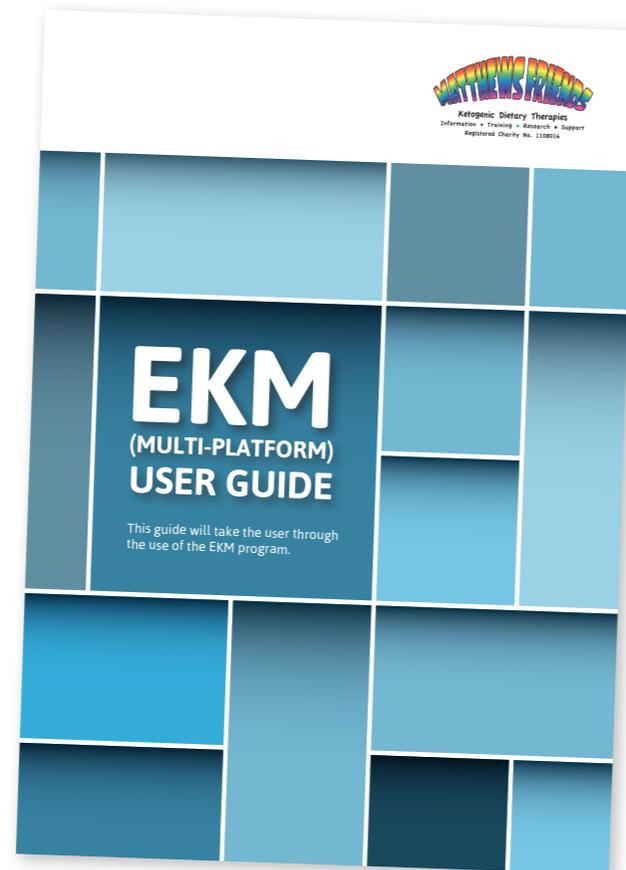
The one condition that Bruce and Liz made when developing this programme was that it would be made available freely to any parent or patient that required it and was medically managed by a ketogenic team, that condition still applies today.

EKM user guide

Bruce Carroll (MicroMan2000 Ltd) worked with Dr. Elizabeth Neal RD, at the Institute of Child Health in London to develop a ketogenic calculator back in the year 2000. This program allows dietitians and those parents and patients that wish to have ability to fine tune or even create meal plans with accurately calculated ratios, fat, protein and carbohydrate intake. It must be stressed that this NOT a substitute for a medically supervised diet.

This programme must be used with the close supervision of a ketogenic dietitian and medical supervision. It is available with all foods from "The Composition of Foods" data set (version 7), however any food can be entered directly into the programme thus EKM can be used with a personal selection of foods or a vast range from the data set provided.

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In Partnership with Matthew's Friends a new phase of development took place in 2016 and a major re-write of the EKM took place which enabled the program to be used within the following operating systems and platforms:

- Mac desktop OS X
- PC, Windows version 7 and above (Vista will work but not recommended)
- Android; phone and tablets (recommend non-Intel platforms)
- iPad and iPhone; iOS

Therefore the majority of desktop and portable devices are now supported by this new release of EKM and the EKM can be taken anywhere as you do not need to be 'online' in order to be able to use it.

Matthew's Friends are delighted to be supporting the EKM and sponsoring its continuing development so as to benefit not only families but also ketogenic dietitians.

For those wishing to download EKM then you will need to confirm that you are being supervised by a ketogenic team.

Please contact ekm@microman2000.co.uk for your EKM access code and registration.

Tutorial films of how to use the EKM are available on the Matthew's Friends website and a User guide is available to download or hard copies can be ordered from Matthew's Friends.

For further information and support please visit the EKM website <http://ekmketocalc.com/>

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New to ketogenic therapy?
Or needing a refresher course
and an opportunity to network
with other ketogenic teams?

This course offers both scientific background and practical training in all aspects of implementation of the different ketogenic therapies. Led by recognised ketogenic diet experts, it includes presentations, practical workshops and time for group discussions. This course has been designed by the Matthew's Friends Medical Advisory Board primarily for new ketogenic teams including dietitians, doctors and other health professionals.

For further information please visit: www.mfclinics.com/ketocollege
or register your interest by emailing: ketocollege@mfclinics.com





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