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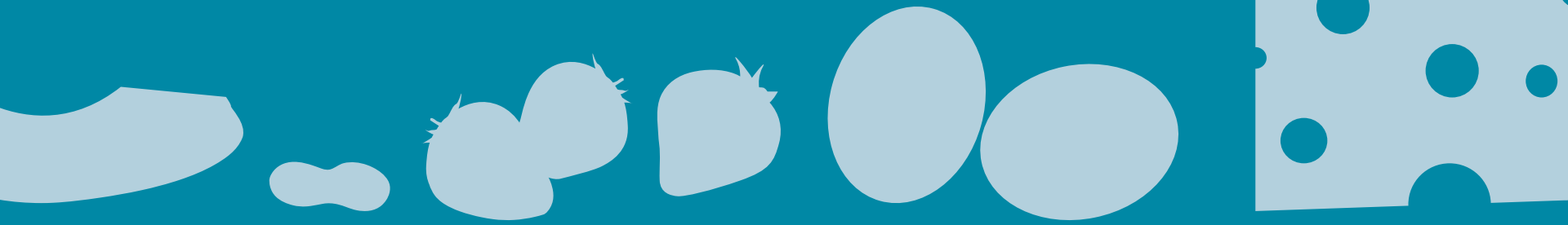
Centre for Education and Development
Endorsed by the BDA for CPD

# An Introduction to Ketogenic Dietary Therapy (KDT) in Complex Epilepsy

We would like to acknowledge the collaboration of MA Leung, B Van de Bor and H Champion in the production of this resource

Updated: May 2021



This information is intended for Health Care Professionals only
BDA endorsement applies only to the educational content of the learning activity

# Contents

# Module 1

## What is Complex Epilepsy and when Should KDT be Considered

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# What is Epilepsy and Management Options?

# What is Epilepsy?

"epilepsy is one of the most common neurological conditions. It affects approximately 600,000 people in the UK[1]"

"electrical activity is happening in the brain all the time as the cells send messages to each other[2]"

"a seizure occurs when there is a sudden burst of intense electrical activity in the brain. This causes a temporary disruption to the way the brain normally works. The result is an epileptic seizure[2]"

**A seizure is an event and epilepsy is the disorder involving recurrent unprovoked seizures**

1. Joint Epilepsy Council (2011) Epilepsy Prevalence, Incidence and Other Statistics; available at http://www.jointepilepsycouncil.org.uk
2. https://www.epilepsy.org.uk/info/what-is-epilepsy

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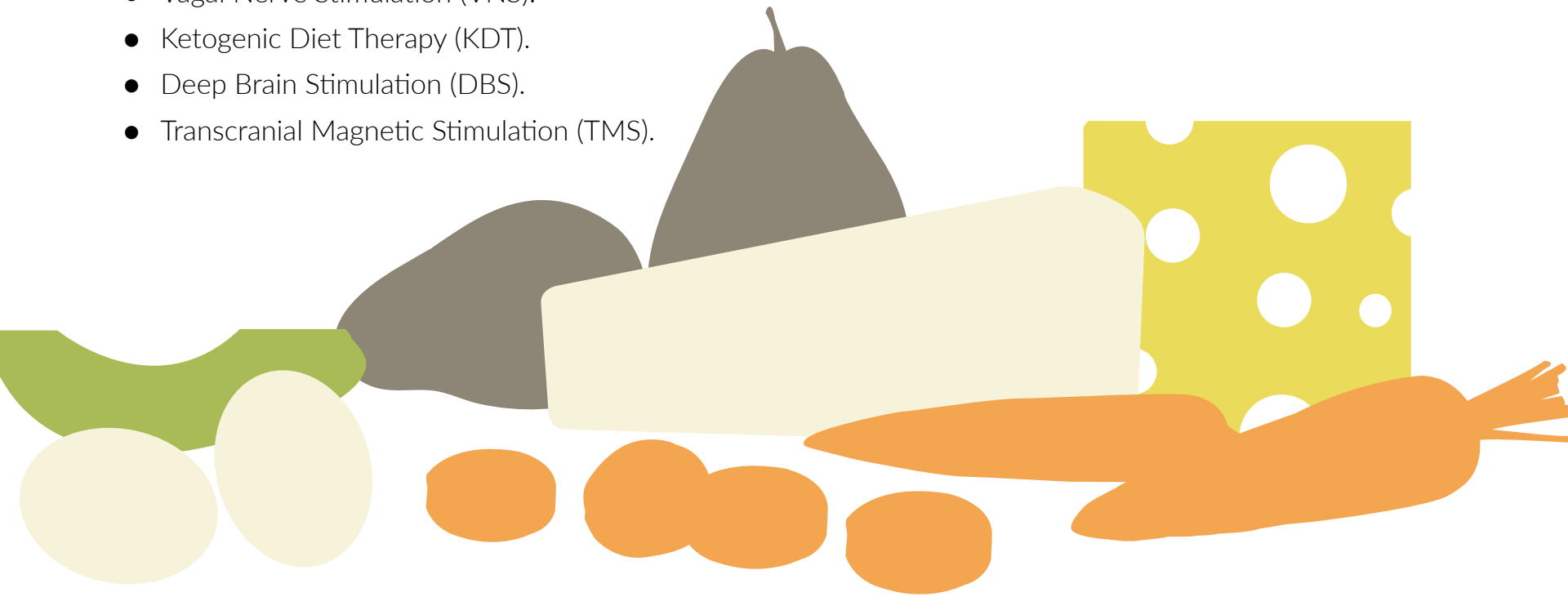
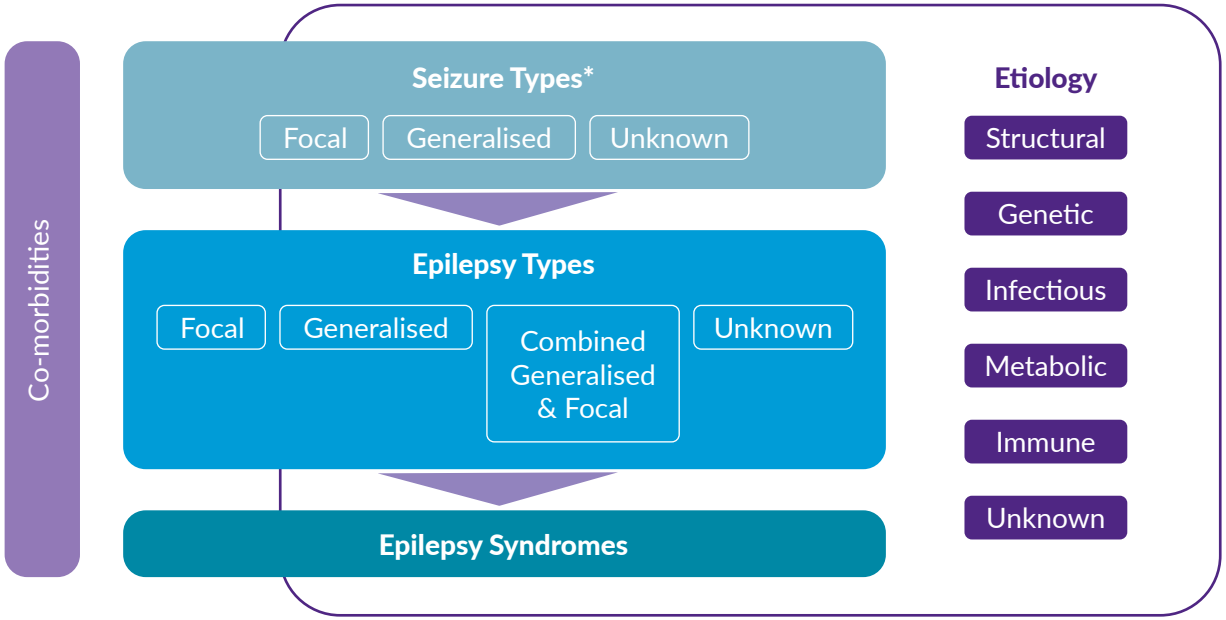
# Epilepsy Management Options

- Anti Seizure Medications (ASMs).
- Epilepsy Surgery.
- Vagal Nerve Stimulation (VNS).
- Ketogenic Diet Therapy (KDT).
- Deep Brain Stimulation (DBS).
- Transcranial Magnetic Stimulation (TMS).



# How to Classify Epilepsy?

**International League Against Epilepsy (ILAE) classification of the epilepsies:**
Position paper of the ILAE Commission for classification and terminology




* *ILAE classification of the epilepsies: Position paper of the ILAE Commission for classification and terminology*, Volume: 58, Issue: 4 Pages 512–521, First published: 08 March 2017, DOI: (10.1111/epl.13709)

# Definition of Epilepsy

**ILAE Official Report:** A Practical Clinical Definition of Epilepsy

**Operational (practical clinical) clinical definition of epilepsy**

## epilepsy

A disease of the brain defined by any of the following conditions

1. At least two unprovoked (or reflex) seizures occurring >24h apart
2. One unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years
3. Diagnosis of an epilepsy syndrome

Epilepsy is considered to be ***resolved*** for individuals who had an age-dependent epilepsy syndrome but are now past the applicable age or those who have remained seizure-free for the last 10 years, with no seizure medicines for the last 5 years.

Epilepsia, 55(4):475–482, 2014 doi: 10.1111/epi.12550

# What is an Epilepsy Syndrome?

If a child is diagnosed with an 'epilepsy syndrome', it means that their epilepsy has some specific signs and symptoms.

**These include:**

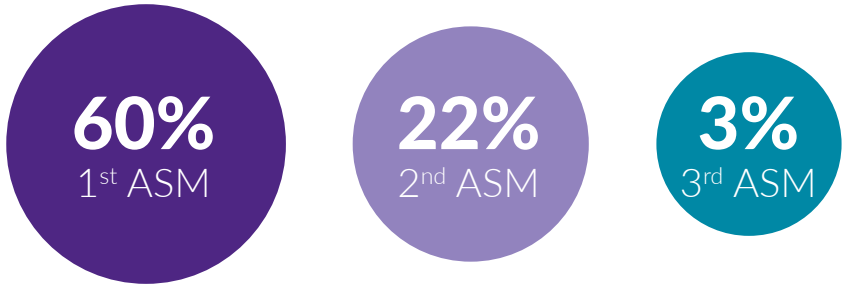
- The type of seizure or seizures they have.
- The age when the seizures first started.
- A specific pattern on an electroencephalogram (EEG).
- Motor problems.

# ASMs in Seizure Control

## Drug-resistant (or refractory) epilepsy is the failure of two or more appropriately chosen ASMs to achieve seizure freedom[1]

- More than 36% of epilepsy patients have inadequate control of seizures with ASMs.[1]
- ASMs are commonly associated with side effects such as drowsiness, blurred vision, dizziness, nausea and vomiting.[2]

**Evidence and clinical consensus show that seizure control diminishes with each successive ASM tried.[3]**




1. Kwan P, Arzimanoglou A, Berg AT et al. Definition of drug resistant epilepsy: consensus proposal by the ad hoc Task Force of the ILAE Commission on Therapeutic Strategies, Epilepsia. 2010 Jun;51(6):1069-77
2. Epilepsy Foundation, 2014. http://www.epilepsy.com/learn/treating-seizures-and-epilepsy/seizure-and-epilepsy-medicines/side-effects. [online] (Accessed 06/01/2015)
3. Wirrell E, Camfield, C, Camfield P et al., Prognostic significance of failure of the initial antiepileptic drug in children with absence epilepsy. Epilepsia, 2001 Jun;42(6):760-3

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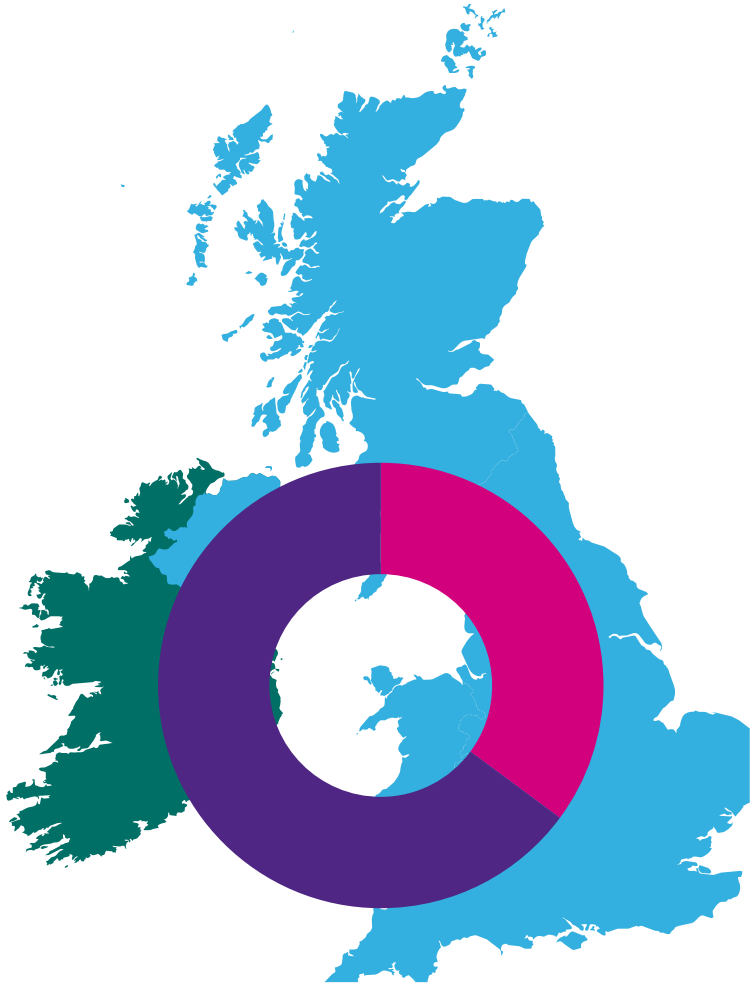
# Resistance to ASMs in the UK[1]

- Population in UK - 64.1M.
- Prevalence of epilepsy - 9.7/1000 (or 1 in 103 people).
- Approximately 600,000 people in the UK have a diagnosis of epilepsy and take anti-seizure medication.

**Drug resistant epilepsy 36%[2] = 210,000**

- 10% are epilepsy surgery candidates
  **= 21,600**
- 90% candidates for other therapies: VNS, ketogenic diet
  **= 194,400**

**Well controlled with anti-seizure medication 64%**



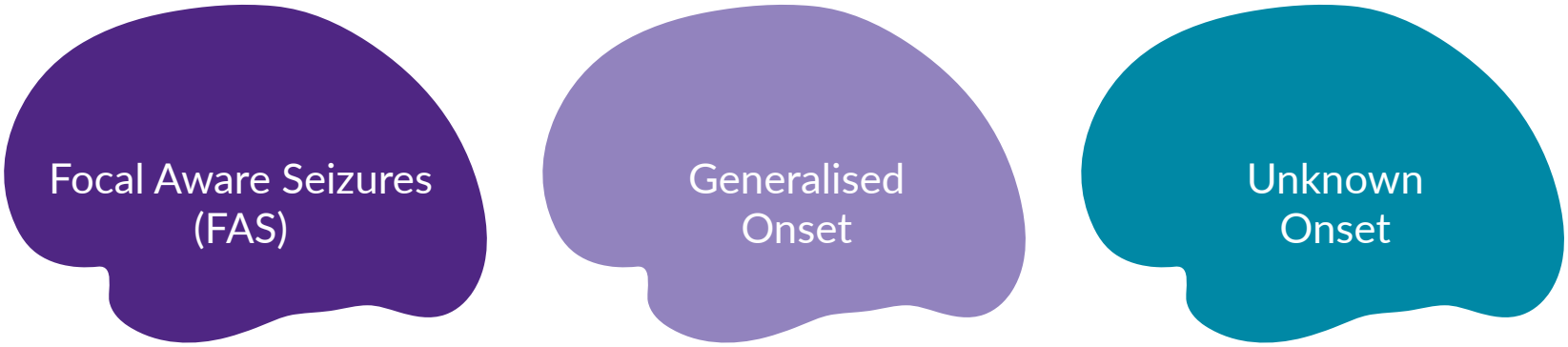
1. Joint Epilepsy Council (2011) Epilepsy prevalence, incidence and other statistics. Joint Epilepsy Council of the UK and Ireland. www.jointepilepsycouncil.org.uk
2. Kwan P, Arzimanoglou A, Berg AT et al. Definition of drug resistant epilepsy: consensus proposal by the ad hoc Task Force of the ILAE Commission on Therapeutic Strategies, Epilepsia. 2010 Jun;51(6):1069-77

# Types of Seizures

# Types of Epilepsy Symptoms

Most individuals equate epilepsy with convulsions, but epileptic seizures can produce many different symptoms. Symptoms may range from whole body convulsions to simply staring into space to barely noticeable muscle twitching.

**3 main groups of seizures are termed:**



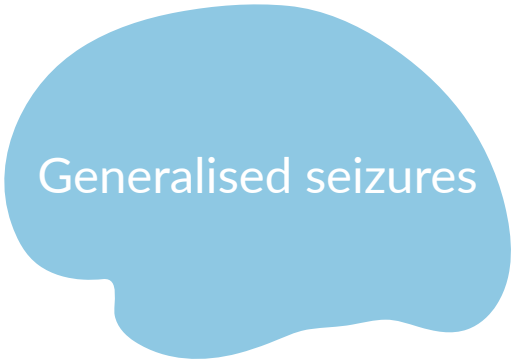
# Focal Aware Seizures (FAS)

- Person is conscious (aware and alert) and will usually know that something is happening/will remember the seizure afterwards.
- During the seizure they may feel ‘strange’ but not able to describe the feeling afterwards. This may be upsetting or frustrating for them.
- FAS are sometimes called ‘warnings’ or ‘auras’ because, for some people, a FAS develops into another type of generalised seizure.

# Generalised Seizures

- Affect both cerebral hemispheres (sides of the brain) from the beginning of the seizure.
- Produce loss of consciousness, either briefly or for a longer period of time, and are sub-categorised into several major types:

1. Tonic clonic (grand mal)
2. Myoclonic
3. Atonic
4. Absence (petit mal)



# Unknown Onset

- This term is sometimes used to describe a seizure if doctors are not sure where in the brain the seizure starts. This may happen if the person was asleep, alone or the seizure was not witnessed.

# Absence Seizure

- Produce symptoms of disconnection from surrounding stimuli.
- Patient appears "absent from their body" and stares off vacantly for a few seconds and then appears normal and has no memory of the incident.

# Tonic Clonic Seizure

**At the start of the seizure:**

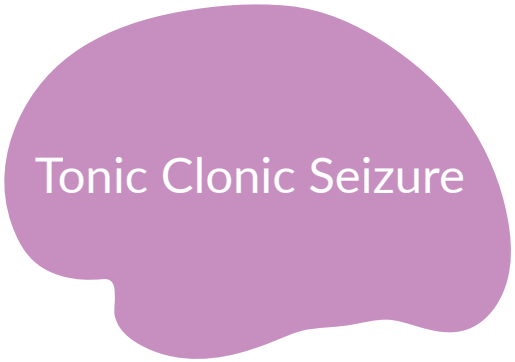
- Unconscious, body goes stiff and may fall backwards.

**During the seizure:**

- Convulsions.
- Breathing might be affected.
- Skin may change.

**After the seizure (once convulsions stops):**

- Breathing and colour return to normal.
- May feel tired, confused, have a headache or want to sleep.



# Myoclonic Seizure

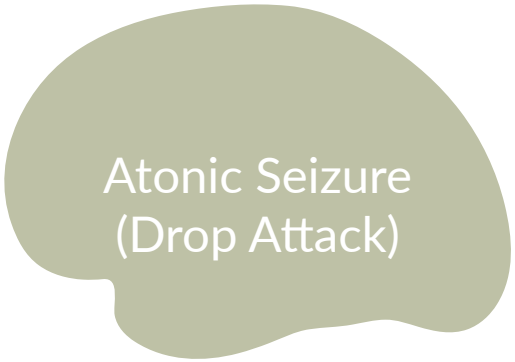
**Myo:** means muscle.

**Clonus:** means rapidly alternating contraction and relaxation of a muscle.

- Usually they don't last more than a second or two.
- There can be just one, but sometimes many will occur within a short time.
- In epilepsy, myoclonic seizures usually cause abnormal movements on both sides of the body at the same time.
- Occur in a variety of epilepsy syndromes that have different characteristics.

# Atonic Seizure (Drop Attack)

- Person's muscles suddenly relax and they become floppy.
- If they are standing they often fall, usually forwards, and may injure the front of their head or face.
- Like tonic seizures, atonic seizures tend to be brief and happen without warning.
- With both tonic and atonic seizures people usually recover quickly, apart from possible injuries.



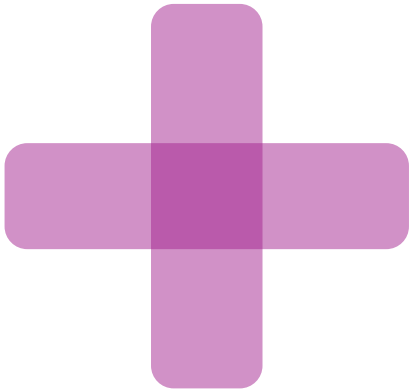
# In Summary

- Epilepsy is expressed in syndromes that describe their features.
- The causes and symptoms of epilepsy are very varied and therefore diagnosis and treatment is extremely complicated.
- Multiple seizure types can be expressed and have an impact on experience of daily life.
- Complex Epilepsy is defined by a resistance to anti-seizure medication.

# Definition of Complex Epilepsy

# What is Complex Epilepsy?

- Intractable 'or uncontrolled' epilepsy is a seizure disorder in which a patient's seizures fail to come under control with medication.
- Various other factors making pharmalogical management very complicated/difficult.
- Drug resistant epilepsy is defined as the failure of adequate trials of two tolerated, appropriately chosen anti-seizure medication.[1]

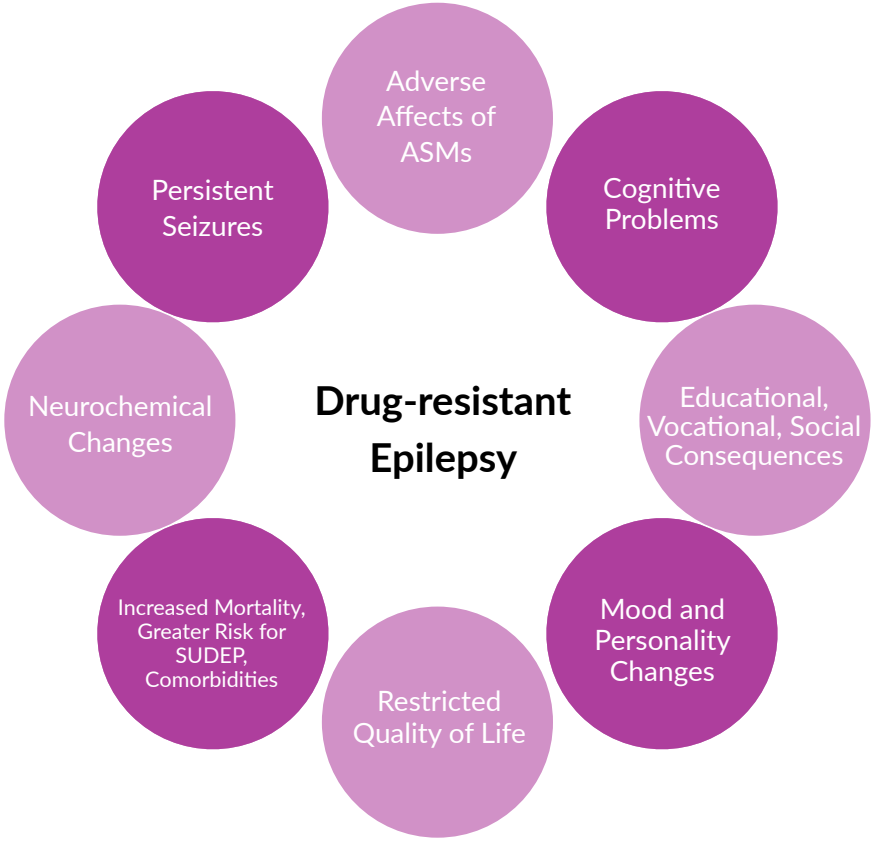


1. Kwan et al Epilepsia. 2010 Jun;51(6):1069-77

# What is Complex Epilepsy?

**Complex because of challenges in:**

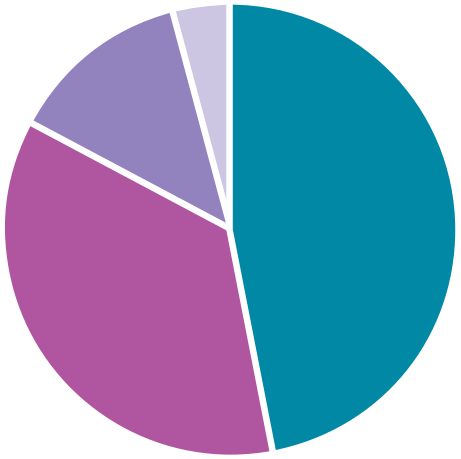
- Diagnosis
- Management
- Adverse affects of ASMs.
- Comorbidities



# Drug-resistant Epilepsy

- 600,000 people in the UK diagnosed with epilepsy and receive anti-seizure medication (ASM); that's 1 in every 103 people.[1]
- Drug-resistant epilepsy is the failure of 2 or more appropriately chosen ASMs to achieve seizure freedom.[2]
- 36% of epilepsy patients have inadequate control of seizures with ASMs.[2]
- Uncontrolled epilepsy can increase the risk of injury, hospital visits, depression, anxiety and SUDEP.[3]
- ASMs are commonly associated with **side-effects** such as drowsiness, blurred vision, dizziness, nausea and vomiting.[4]

## Chances of ASM success diminishes after every failure[5]



N=470 previously untreated epilepsy patients treated with ASMs

- **47%** seizure free 1st ASM
- **13%** seizure free 2nd ASM
- **4%** seizure free 3rd or multiple ASMs
- **36%** Not seizure free

Drug-resistant epilepsy may require an alternative management option


1. Joint Epilepsy Council (2011) 'Epilepsy Prevalence, Incidence and Other Statistics', Available here (Accessed: 16th April 2018).
2. Kwan, P., Arzimanoglou, A. and Berg, A (2010) 'Definition of Drug-resistant Epilepsy: Consensus Proposal by the Ad Hoc Task Force of the ILAE Commission on Therapeutic Strategies', Epilepsia, 51(6), pp. 1069–1077
3. Epilepsy Society (2018) 'Risks with Epilepsy', Available here (Accessed: 18th September 2018).
4. Epilepsy Foundation (2018) 'Risks with Epilepsy', Available here (Accessed: 18th September 2018).
5. Kwan, P. and Brodie, M.J (2000) 'Early identification of refractory epilepsy', New England Journal of Medicine, 342(5), pp. 314–319.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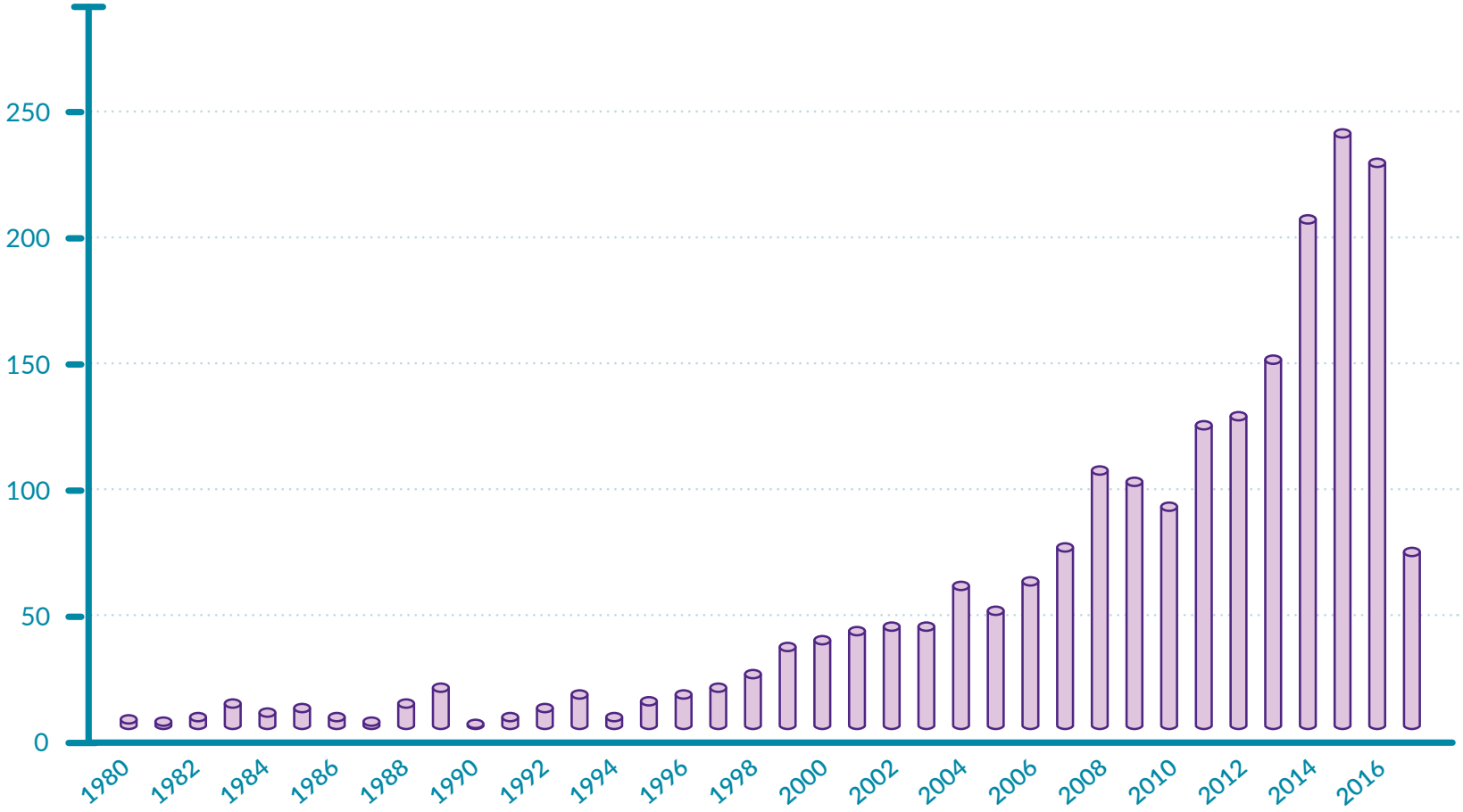
# Publications by Year



# Efficacy of KDT in Complex Epilepsy

# Efficacy of the Ketogenic Diet[1]

- A heritage of clinical evidence shows the efficacy of the ketogenic diet.
- More than 50% of patients experience over 50% reduction in seizures after starting a ketogenic diet.

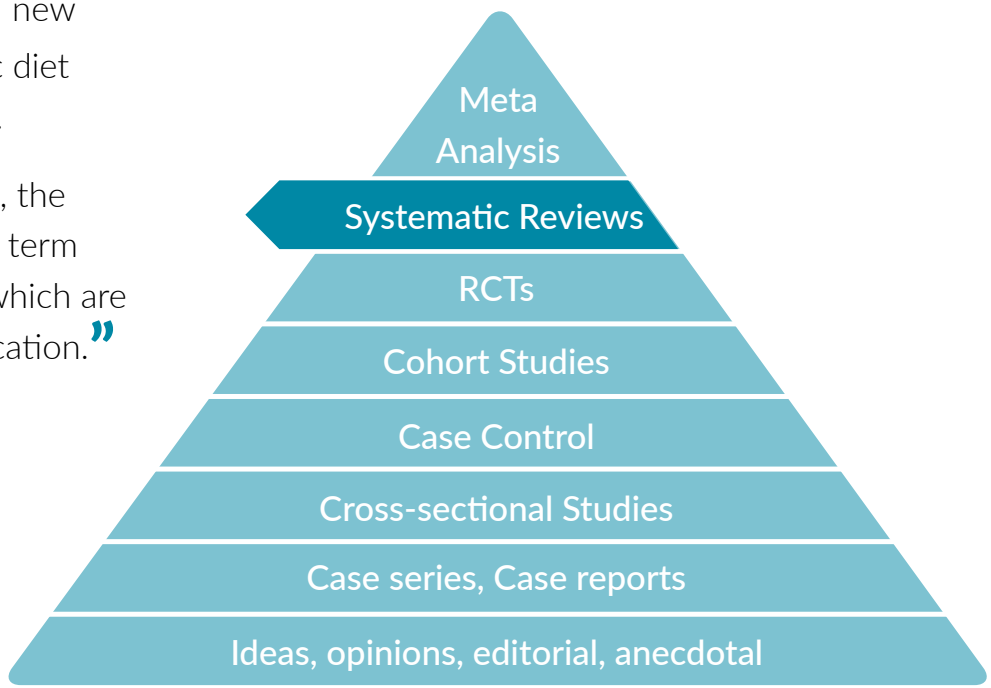
1. Martin-McGill KJ et al. Ketogenic diets for drug-resistant epilepsy. Cochrane Database of Systematic Reviews 2020, Issue 6. Art. No.: CD001903. DOI: 10.1002/14651858.CD001903.pub5

# Efficacy of the Ketogenic Diet[1]

**The Cochrane review 2020 states[1]:**

"Our review update included data from 5 new randomised clinical trials of the ketogenic diet randomised studies of the ketogenic diet.

(...) These studies suggest that in children, the ketogenic diet results in short to medium term benefits in seizure control, the effect of which are comparable to modern anti seizure medication."




1. Martin-McGill KJ et al. Ketogenic diets for drug-resistant epilepsy. Cochrane Database of Systematic Reviews 2020, Issue 6. Art. No.: CD001903. DOI: 10.1002/14651858.CD001903.pub5

# Efficacy of the Ketogenic Diet

**The Cochrane review 2020 states[1]:**

"The randomised controlled trials discussed in this review show promising results for the use of KDTs in epilepsy.

For people who have drug resistant epilepsy or people who are not suitable for surgical intervention, a KDT remains a valid option; (...)"


1. Martin-McGill KJ et al. Ketogenic diets for drug-resistant epilepsy. Cochrane Database of Systematic Reviews 2020, Issue 6. Art. No.: CD001903. DOI: 10.1002/14651858.CD001903.pub5

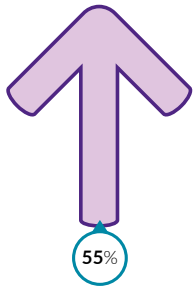
# Efficacy of the Ketogenic Diet[1]

More than 50% of patients experience over 50% reduction in seizures after starting a ketogenic diet.[1]

- Review of efficacy based on randomised control trials.
- The Cochrane Collaboration review included 13 RCTs, resulting in 12 publications.
- All applied an intention-to-treat analysis.
- A total of 932 participants were recruited; 711 children (4 months to 18 years) and 221 adults (16 years and over).
- No meta-analysis due to heterogeneity of trials.

**Reported rates in children after 3 months on:**

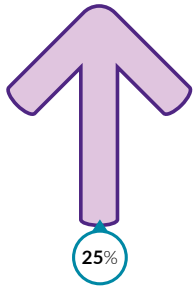
Classical Ketogenic Diet:



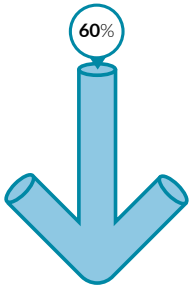

**Seizure freedom** rates reached up to **55%**


**Seizure reduction** rates reached up to **85%**

Modfied Atkins Diet (MAD)




**Seizure freedom** rates reached up to **25%**




**Seizure reduction** rates reached up to **60%**

1. Martin-McGill KJ et al. Ketogenic diets for drug-resistant epilepsy. Cochrane Database of Systematic Reviews 2020, Issue 6. Art. No.: CD001903. DOI: 10.1002/14651858.CD001903.pub5

# Review of Efficacy Based on Non-randomised Controlled Trials

In a systematic review by Keene, intention to treat analysis, patients on KDT after 6 months presented with[1]:

**15.6%** complete seizure freedom (95% CI 10.4-20.8)

**33%** greater than 50% seizure reduction (95% CI 24.3-41.8)

In a meta-analysis by Henderson, those patients that were compliant to the KDT showed[2]:

**24%** complete seizure freedom

**52%** greater than 90% seizure reduction

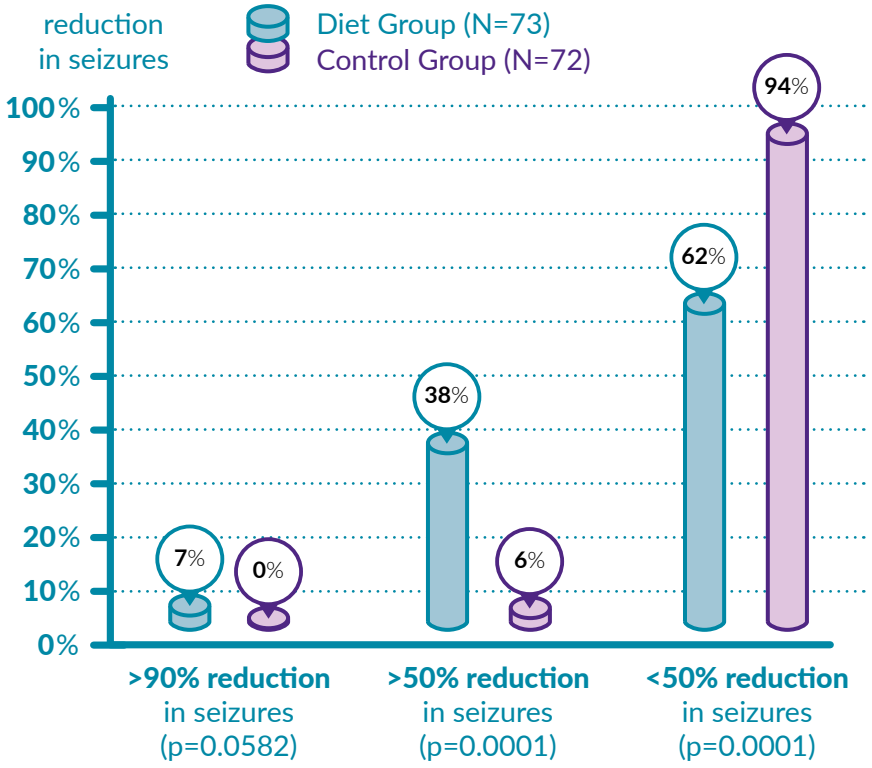
1. Keene D. Pediatric Neurology 2006;35(1):1-5
2. Henderson C et al. J Child Neurol 2006;21:193-8

# The Ketogenic Diet for the Dietary Management of Childhood Epilepsy

**A randomised controlled Trial[1]**

This graph shows the number of children in each group who achieved 50% and 90% seizure reduction at 3 months.

| | Data from 145 children recruited between 2–16 years old | |
|---|---|---|
| Randomised assigned to | 73 children on KDT | 72 controls |




1. Neal NG et al. Lancet Neurol 2008;7:500-6

# Implementing the Ketogenic Diet Early has Beneficial Results

- Many seizures before therapy, as well as inadequate response to initial drug treatment, with less than three previous ASMs are positive predictive factors of drug resistant epilepsy.[1]
- Starting the ketogenic diet at a lower age means seizure freedom is more often achieved and maintained in infants.[2]
- Shorter duration of epilepsy independently predicts higher seizure freedom rates.[5,6,7,8,9]
- Shorter duration of epilepsy is related to higher IQ.[3,4,5,6]


1. Kwan P et al. N Engl J Med 2000;342:314-9
2. Dressler A et al. Epilepsy Res 2015;116:53-8
3. Jonas R et al. Neurology 2004;62(10):1712-21
4. Basheer S et al. Epilpesia 2007;48(1):133-40
5. Thomas X et al. Int J Pediatrics 2012; DOI 1155/2012/527891
6. D'Argenzio L et al. Epilepsia 2011;52(11):1966-72
7. Englot DJ et al. J Neurosurg 2011;115(6):1248-55
8. Simasathien T et al. Ann Neurol 2013;73(5):646-54
9. Lamberink H et al. Epileptic Disord 2015;17(3):211-28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# The Use of KDT in Infants

**Evidence of efficacy**

- Syndromes of early childhood epilepsy are difficult to manage and associated with significant morbidity and mortality.[1]
- Neonates and infants produce and utilise ketone bodies as well as older children.[2]
- KDT is increasingly being used in catastrophic epilepsies in infancy and young children.[3]
- KDT is a safe and effective management option for infants and young children with refractory epilepsy.[4]
- KetoCal 3:1 is a specialist ketogenic formula suitable for infants.


1. Rubenstein JE. Epilepsia 2008;49(8):30-2
2. Wheless JW. Nonpharmacologic treatment of the catastrophic epilepsies of childhood. Epilepsia. 2004;45 suppl5:17-22
3. Kossoff EH et al. Epilepsia 2008;49(8):37–41
4. Nordli DR jr et al. Experience with the ketogenic diet in infants. Pediatrics 2001;108:129-133

# The Use of KDT in Adults

- Approximately half of all adults that commence KDT can expect a reduction of at least 50% in seizures.[1,3]
- Reports in adults with epilepsy indicate KDT can[2]:
  - Improve energy levels.
  - Increase clarity of thought.
  - Accelerate recovery from seizures.
  - Improve quality of life.
- Modified KDT is often tolerated better in adolescent and adults and adherence can be high where seizure reduction benefits are seen.[2]
- Supplementing modified KDT with ketogenic formula significantly increases long term compliance in adults with drug-resistant epilepsy.[3]

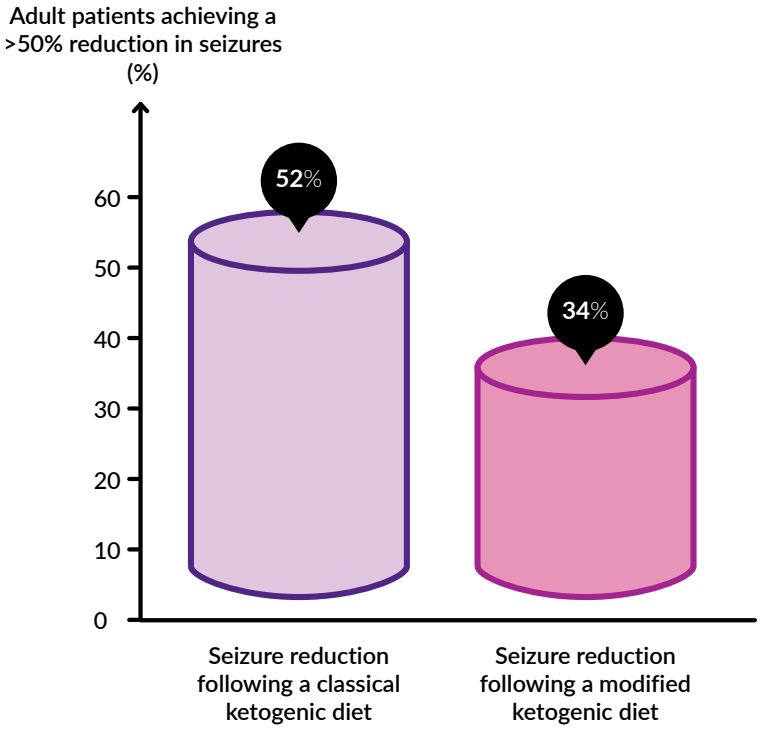
1. Kossoff EH et al. Epilepsia 2008;49(8):37–41
2. Sirven J, Whedon B, Caplan D, Liporace J, Glosser D, O'Dwyer J, Sperling MR. 1999 The ketogenic diet for intractable epilepsy in adults: Preliminary results. Epilepsia, 40(12):1721-1726
3. Mcdonald, TJW et al. (2018) Improving compliance in adults with epeulspy on a modified atkins diet: a randomised trial.Seizure, 60, 132-138

# The Use of KDT in Adults[1]

## EFFICACY FOR KDT IN ADOLESCENTS AND ADULTS

Meta-analysis indicating that KDT is an effective treatment in adults with drug-resistant epilepsy (n=270).

- Success rates up to 70% have been reported in adults using KDT.[1]
- Approximately half of all adolescents and adults that commence KDT can expect at least a 50% reduction in seizures.[2,3]
- Modified KDT is often tolerated better in adults and adherence can be high where seizure reduction benefits are seen.[4]




1. *Fang, Y., Xiao-Jai, L. et al. (2015) 'Efficacy of and Patient Compliance with a Ketogenic Diet in Adults with Intractable Epilepsy: A Meta-Analysis', Journal of Clinical Neurology, 11(1), pp.26–31.*
2. *Kossoff, E.H. and Dorward, J.L. (2008) 'The Modified Atkins Diet', Epilepsia, 49(8), pp.37–41.*
3. *McDonald, T.J.W., Henry-Barron, B.J. et al (2018) 'Improving Compliance in Adults with Epilepsy on a Modified Atkins Diet: A Randomized Trial', Seizure, 60(10), pp.132–138.*
4. *Sirven, J. and Whedon, B et al. (1999) 'The Ketogenic Diet for Intractable Epilepsy in Adults: Preliminary Results', Epilepsia, 40(12), pp.1721–1726.*

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# Efficacy of the Ketogenic Diet as a Management Option for Epilepsy: Meta Analysis

**In meta-analysis by Henderson, those patients that were compliant to the KDT showed[1]:**

**24%** Complete seizure freedom.

**52%** Greater than 90% seizure reduction.

**2006:**

- 19 studies, 1084 patients.

**Conclusion:**

- 84% responded with at least a 50% reduction in seizures.
- 1 in 4 experienced complete seizure freedom.

1. Efficacy of the ketogenic diet as a treatment option for epilepsy: meta-analysis. Henderson CB et al J Child Neurol. 2006 Mar;21(3):193-8

# Updated Guidelines

- In 2012, NICE updated their Clinical Guideline [CG 137].
- Epilepsies: diagnosis and management.

NICE National Institute for Health and Care Excellence

# Nice Guideline

- Refer children and young people with epilepsy whose seizures have not responded to appropriate ASMs to a tertiary paediatric epilepsy specialist for consideration of the use of a ketogenic diet[1].
- View the NICE Guideline here.


1. NICE Epilepsies: diagnosis and management. Clinical guideline [CG137] Published date: January 2012 Last updated: February 2016

# When to Refer to a Tertiary Centre

- If seizures are not controlled and/or there is diagnostic uncertainty or treatment failure, refer to tertiary services soon for further assessment. Consider referral when one or more of the following criteria are present:
- The epilepsy is not controlled with medication within 2 years.
- Management is unsuccessful after 2 or more appropriately prescribed ASMs.
- The child is aged under 2 years.
- A child, young person or adult experiences, or is at risk of, unacceptable side effects from medication.
- There is a unilateral structural lesion.
- There is psychological and/or psychiatric comorbidity.
- There is diagnostic doubt as to the nature of the seizures and/or seizure syndrome.

# International Consensus Guidelines for the Management of Children on KDT

## First published in 2009 and recently updated in 2018[1]

**Eric H. Kossoff is a professor of neurology and paediatrics at Johns Hopkins Hospital in Baltimore**

**Summary**

Ketogenic dietary therapies (KDT's) are established, effective nonpharmacologic treatments for intractable childhood epilepsy. For many years KDT's were implemented differently throughout the world due to lack of consistent protocols. In 2009, an expert consensus guideline for the management of children on KDT was published, focusing on topics of patient selection, pre-KDT counselling and evaluation, diet choice and attributes, implementation, supplementation follow-up, side events, and KDT discontinuation. It has been helpful in outlining a state-of-the-art protocol, standardizing KDT for multicenter clinical trials, and identifying areas of controversy and uncertainty for future research.

Now one decade later, the organizers and authors of this guideline present a revised version with additional authors, in order to include recent research, especially regarding other dietary treatments, clarifying indications for use, side effects during initiation and ongoing use, value of supplements and methods of KDT discontinuation. In addition, authors completed a survey of their institution's practices, which was compared to responses from the original consensus survey, to show trends in management over the last 10 years.

Kossoff E. H. et al (2018) 'Optimal clinical management of children receiving dietary therapies for epilepsy: Updated recommendations of the International Ketogenic Diet Study Group' Epilepsia Open, 3(2):175–192, doi: 10.1002/epi4.12225

# Changes in Indications

**2009**

**Table 1[1]**
Epilepsy Syndromes and conditions in which KDT has been reported as particularly beneficial

**Probable Benefit (at least two publications)**
Glucose transporter protein I (GLUT-I) deficiency
Pyruvate dehydrogenase deficiency (PDHD)
Myoclonic-astatic epilepsy (Doose syndrome)
Tuberous sclerosis complex
Rett syndrome
Sever myoclonic epilepsy of infancy (Dravet syndrome)
Infantile spasms
Children receiving only formula (infants or enterally fed patients)

**Suggestion of benefit (one case report or series)**
Selected mitochondrial disorders
Glycogenosis type V
Landau-Kleffner syndrome
Lafora body disease
Subacute sclerosing panencephalitis (SSPE)

**2018**

**Table 1[2]**
Epilepsy Syndromes and conditions (listed alphabetically) for which KDT has been consistently reported as more beneficial (>70%) that the average 50% KDT response (defined as >50% seizure reduction).

**Angelman syndrome**
**Complex I mitochondrial disorders**
Dravet syndrome
Epilepsy with myoclonic-atonic seizures (Doose syndrome)
Glucose transporter protein I (Glut-I) deficiency syndrome (Glut IDS)
**Febrile infection-related epilepsy syndrome (FIRES)**
Formula-fed (solely) children or infants
Infantile spasms
**Ohtahara syndrome**
Pyruvate dehydrogenase deficiency (PDHD)
**Super-refractory status epilepticus**
Tuberous sclerosis complex

1. Kossoff, E. H. (2008), International consensus statement on clinical implementation of the ketogenic diet: Agreement, flexibility, and controversy. Epilepsia, 49: 11-13. doi:10.1111/j.1528-1167.2008.01823.x
2. Kossoff E. H. et al (2018) 'Optimal clinical management of children receiving dietary therapies for epilepsy: Updated recommendations of the International Ketogenic Diet Study Group' Epilepsia Open, 3(2):175–192, doi: 10.1002/epi4.1222

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Lafora body disease
Subacute sclerosing panencephalitis (SSPE)

**2018**

**Table 1[2]**
Epilepsy Syndromes and conditions (listed alphabetically) for which KDT has been reported as moderately beneficial (not better than the average dietary therapy response, or in limited single-center case reports).

**Angelman syndrome**
**CDKL5 encephalopathy**
**Childhood absence epilepsy**
**Cortical malformations**
**Epilepsy of infancy with migrating focal seizures**
**Epileptic encephalopathy with continuous spike-and-wave during sleep**
Glycogenosis type V
Juvenile myoclonic epilepsy
Lafora body disease
Landau-Kleffner syndrome
**Lennox-Gastaut syndrome**
**Phosphofructokinase deficiency**
Rett syndrome
Subacute sclerosing panencephalitis (SSPE)

1. Kossoff, E. H. (2008), International consensus statement on clinical implementation of the ketogenic diet: Agreement, flexibility, and controversy. Epilepsia, 49: 11-13. doi:10.1111/j.1528-1167.2008.01823.x
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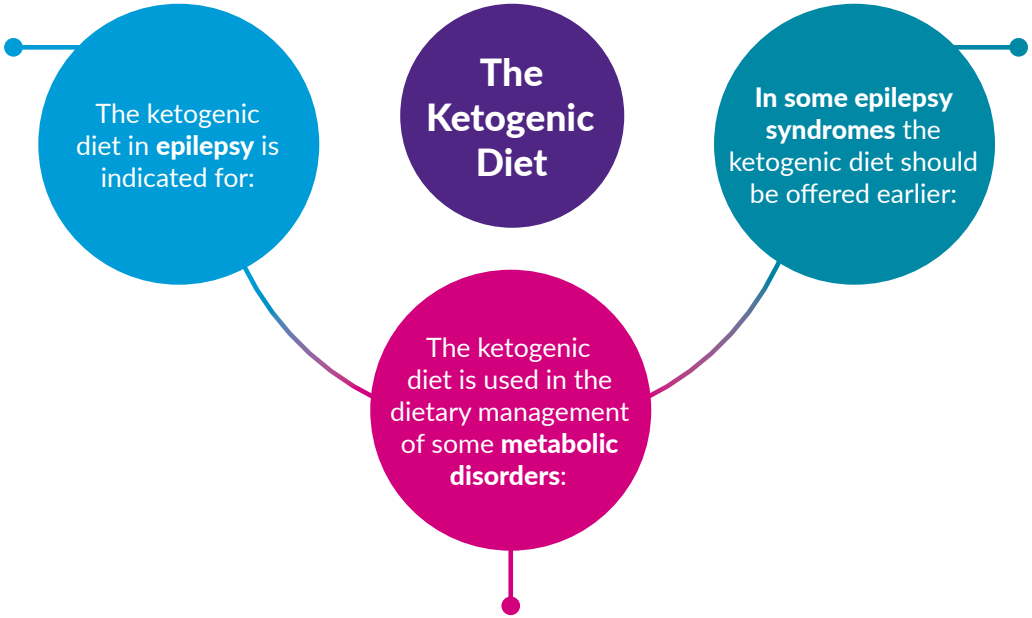
# Patients who Benefit from the Ketogenic Diet[1]

- Patients who have failed two to three ASMs
- Patients who do not tolerate and/or experience severe side effects from ASMs
- Epilepsy patients with gastrostomy feeding tubes



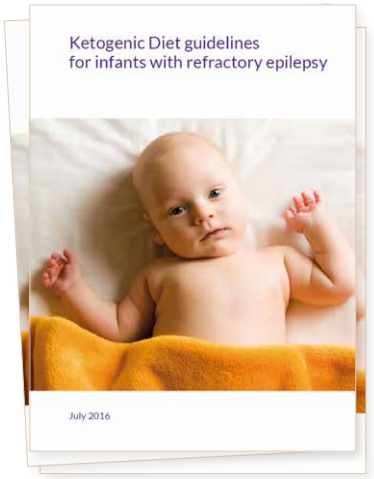

- Infantile spasms
- Lennox-Gastaut Syndrome
- Dravet Syndrome
- Doose Syndrome (myoclonic-astatic epilepsy)
- Status epilepticus
- FIRES (febrile infection-related epilepsy syndrome)
- Angelman syndrome
- Ohtahara syndrome
- Tuberous sclerosis complex

- Glucose 1 transporter (GLUT 1) deficiency
- Pyruvate dehydrogenase (PDH) deficiency

1. Kossoff, E. et al., 2018 Optimal Clinical Management of Children Receiving Dietary Therapies for Epilepsy: Updated Recommendations of the International Ketogenic Diet Study Group. Epilepsia Open: 1-18,

# Guidelines for Infants with Epilepsy[1]

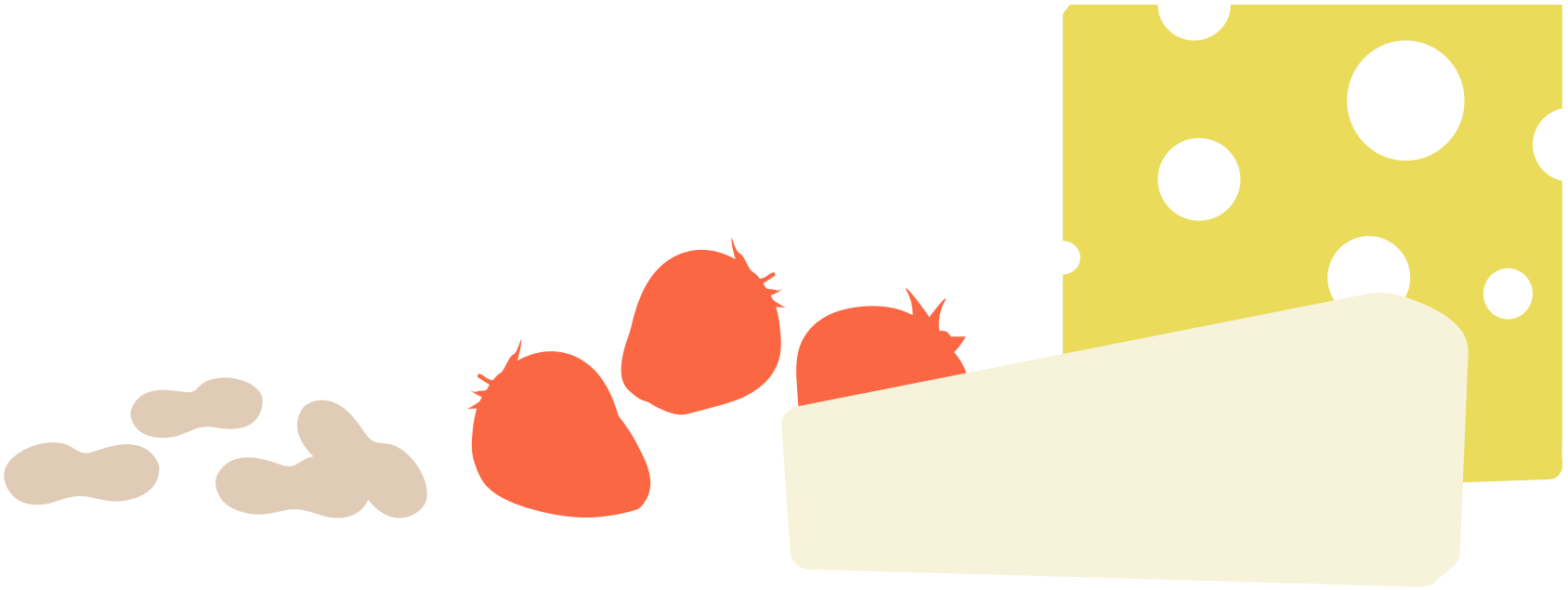
- Ketogenic diet management in infants with refractory epilepsy is effective and can be done safely.
- Ketogenic diet must be adjusted to specific nutritional requirements of infants.




1. Van der Louw E et al European J Paediatr Neurol. 2016 Nov;20(6):798-809

# So in Summary

- KDT has proven efficacy in children and infants, and a growing body of evidence supporting its use in adults.



# When Should KDT be Considered?

# When Should Ketogenic Dietary Therapy (KDT) be Considered?

National Institute of Health and Care Excellence (NICE) in 2012 and updated in 2016 recommended that after the failure of 2 or more appropriately prescribed ASMs alternative therapies should be considered[1].



1. NICE Epilepsies: diagnosis and management. Clinical guideline [CG137] Published date: January 2012 Last updated: February 2016

# When Should KDT be Considered

As a third or fourth line management option, KDT is more likely to improve seizure control and the impact of seizures on daily life than the introduction of subsequent ASMs.

Children who are solely fed via a tube, gastrically or jejunely  will find changing their feed to a ketogenic feed requires very little change to the home enteral feeding routine.

# Module 2

## How Does KDT Work, Possible Side Effects and How They Are Managed

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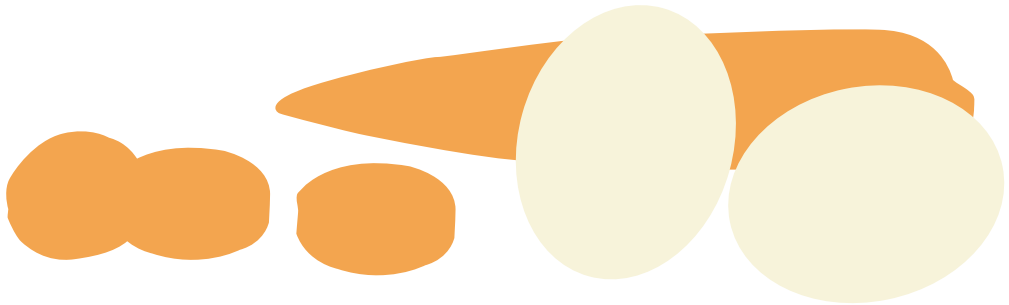
# Types of KDT

# Types of Ketogenic Dietary Therapy

There are 4 different versions of ketogenic diets. All have restricted carbohydrate, are rich in fats and provide sufficient protein to support growth and development. Research has shown that all versions of the diet are effective in reducing seizures[1].



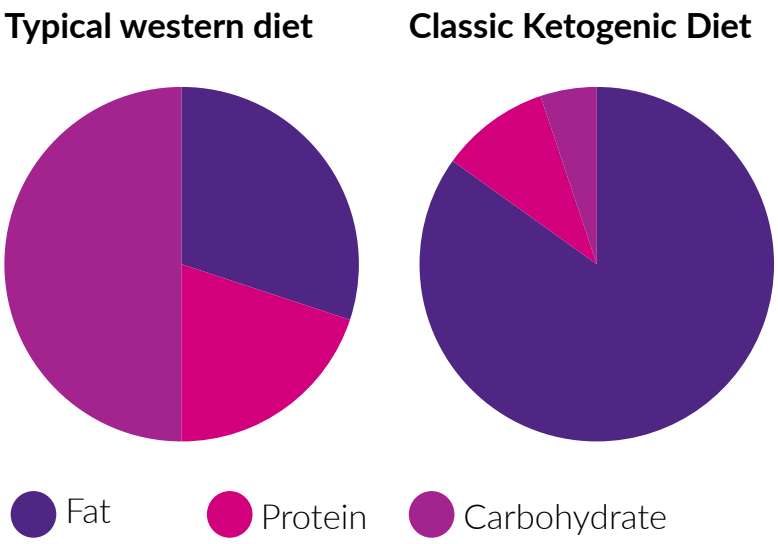
1. Martin-McGill KJ et al. Ketogenic diets for drug-resistant epilepsy. Cochrane Database of Systematic Reviews 2020, Issue 6. Art. No.: CD001903. DOI: 10.1002/14651858.CD001903.pub5

# The Classical Diet

**2:1 – 4:1**

First described in the 1920's, is based on a ratio of fat to protein and carbohydrate. Ratios typically are between 2:1 to 4:1. Ratios are flexible and tailored to a patients needs by a dietitian.

The classical diet is suited to infants or enterally fed patients, and those who have limited food choices in their diet.




# Ketogenic Diet: Classical

- Very high fat diet, often between 80-90% of total energy.
- calculated on a ratio of fat to carbohydrate and protein combined. e.g. 4:1 ratio equates to 4g of fat to every 1g of protein and carbohydrate combined.
- In terms of energy, this means that 90% of calories comes from fat and remaining 10% from protein and carbohydrate combined.
- Ratio can be adjusted, depending on level of ketosis and effect on seizure management.
- Older children and adults readily require lower ratios due to their higher protein requirements

# Ketogenic Diet: Classical

- Currently the most widely used of the ketogenic diets with the largest amount of published research.
- Used since the 1920s in children before the widespread use of ASMs.[1]

1. Neal EG, Chaffe H, Schwartz RH, Lawson MS, Edwards N, Fitzsimmons G, et al. The ketogenic diet for the treatment of childhood epilepsy: a randomised controlled trial. Lancet Neurology. 2008;7(6):500–6.

# The Medium Chain Triglyceride Diet (MCT)

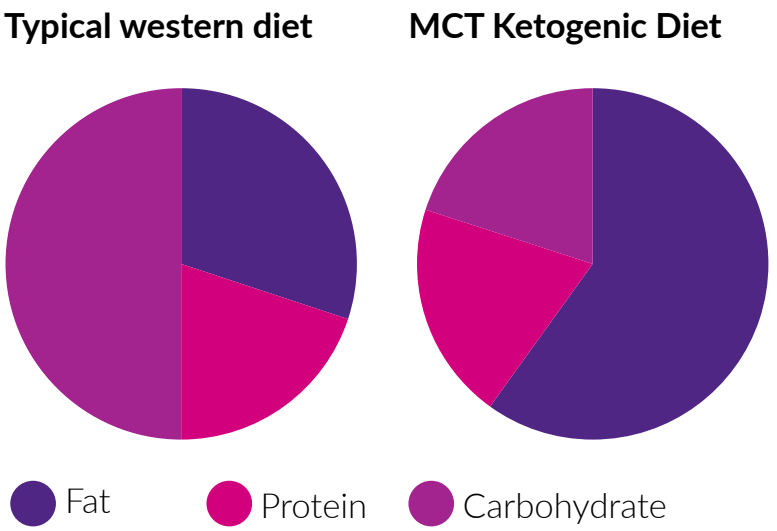
MCT

Medium chain triglycerides are metabolised to ketones with little metabolic regulation. MCT ketogenic diets allow a more generous quantity of carbohydrate to be included in the daily diet. MCT is provided as a prescribed food supplement that can be added to milk or mixed into food.

School age children manage MCT supplements well and enjoy the flexibility of the additional carbohydrate allowance.

## KETOGENIC DIET: MCT

- 75% energy from fat with typically 45% of the fat as MCT fat.
- The remaining calories are derived from a combination of protein and carbohydrate.
- No ratio calculations are required.



# Ketogenic Diet: MCT

MCT

- 30-60% of the energy is derived from the supplementation of MCT fat, such as Liquigen or MCT Oil.
- The remainder of dietary energy is derived from LCT fat, protein and carbohydrate.

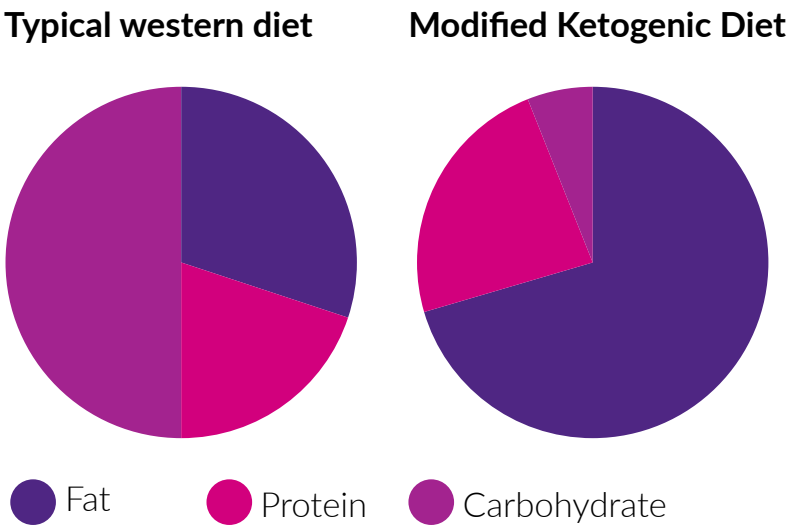
Liquigen and MCT Oil are Foods for Special Medical Purposes for the dietary management of patients with fat malabsorption and other proven malabsorption syndromes and must be used under medical supervision.

# Modified Ketogenic Diet (MKD)

MKD

Based on the same principle of the Atkins Diet for weight loss, but without an energy deficit. Energy equilibrium is maintained with the addition of fat to the diet. The diet focuses strongly on carbohydrate restriction while allowing protein foods to be eaten more liberally.

This Ketogenic diet, with its flexibility on protein is a popular choice for older children and young adults. Less preparation and planning is required making it easier to eat away from home.



# Ketogenic Diet: Modified

MKD

- This diet is managed without protein or calorie restriction alongside high fat and low carbohydrate food choices.
- Origin of diet is an adaption of the Atkins Diet used for weight loss. In the USA termed The Modified Atkins Diet (MAD) Suitable for older children and adolescents.
- Fits into independent lifestyles.
- Household measures are used rather than weighing of foods.
- Carbohydrate intake restricted to 10-30g per day.

# Low Glycaemic Index Treatment (LGIT)

The LGIT focuses on both the amount and type of carbohydrate allowed each day. The diet is high in fat like ketogenic diets.

The GI of carbohydrate is a measure of how readily the carbohydrate raises blood sugar levels. Carbohydrates with a low GI are allowed in quite generous quantities. These carbohydrates are usually associated with fibre.

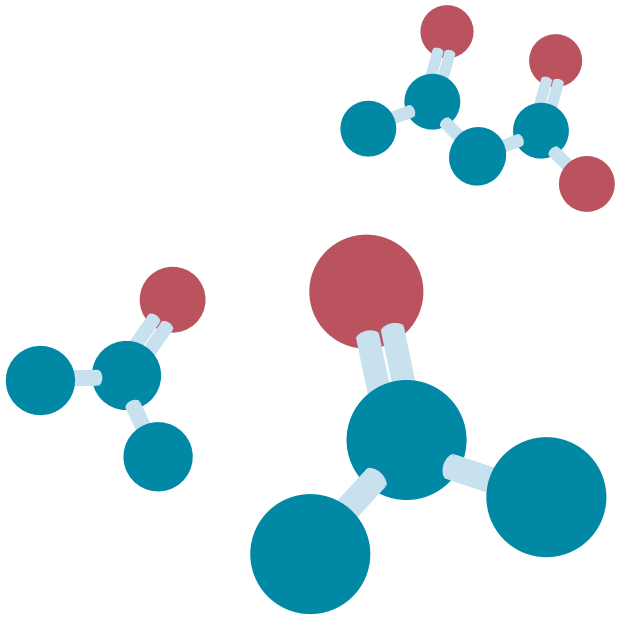
This diet or option is useful for older children. The high fibre content can be a challenge for younger children.

# How Does KDT Work?

# How Does Ketogenic Diet Therapy Work?

Like ASMs, it is likely there is no single mechanism to explain the effect of KDT in this heterogeneous group of neurological disorders.

Prominent shifts in metabolism seem to underlie the effects of KDT and ketone bodies themselves may have a disease modifying effect through epigenetics.



(Ketones)

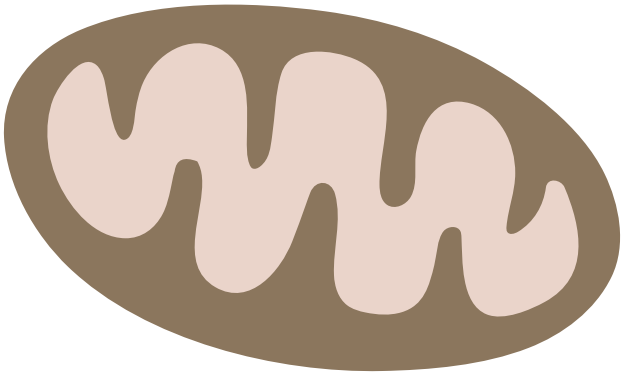
# Known Mechanisms of Action:

**Direct effect of fuel source for brain function**
Ketones cross the blood brain barrier increasing energy available to the brain.

Reduction in glycolysis (due to reduced carbohydrate diet) having a direct anticonvulsant effect.

**Increase in energy production in brain tissue**
Mitochondrial Biogenesis; Decanoic acid; C10 has been linked to proliferation of active mitochondria in brain tissue[1].

Research has shown that decanoic acid is produced as a result of the ketogenic diet. This helps reduce seizures in some people[2].



(Mitochondrial Biogenesis)

1. Link to Hughes et al J neurochem 2014 129(3): 426-33g
2. Chang et al Brain, Volume 139, Issue 2, 1 February 2016, Pages 431–443

# Known Mechanisms of Action:

**Alteration to Neurotranmitters Metabolism[1]**

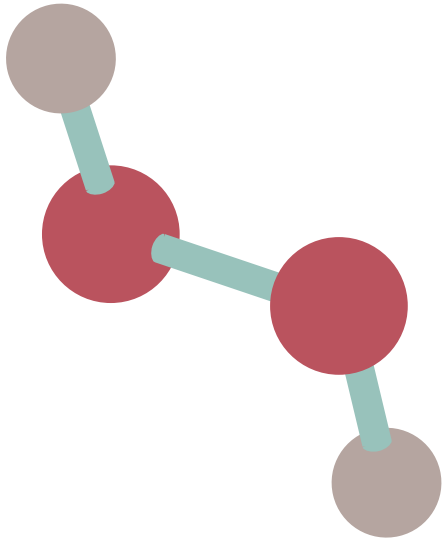
GABA (gamma-aminobutyric acid), an inhibitory neurotransmitter is increased in the presence of ketone bodies. The mode of action is through:

**Reduction of Oxidative Stress[2]**

Ketones metabolism reduces mitochondrial production of $H_2O_2$. This reduces oxidative stress.

**Epigenetic Mechanisms[3]**

Ketone bodies bind to genes changing their expression.



($H_2O_2$ Hydrogen Peroxide)

1. Hartman AL, Gasior M, Vining EP, Rogawski MA. The neuropharmacology of the ketogenic diet. Pediatr Neurol. 2007;36(5):281–292. doi:10.1016/j.pediatrneurol.2007.02.008
2. Shimazu et al Suppression of oxidative stress by betahydroxybutyrate an histone deacetylase inhibitor. Science. 2013 Jan 11;339(6116):211-4
3. Newman JC, Verdin E. Ketone bodies as signaling metabolites. Trends Endocrinol Metab. 2014;25(1):42–52. doi:10.1016/j.tem.2013.09.002

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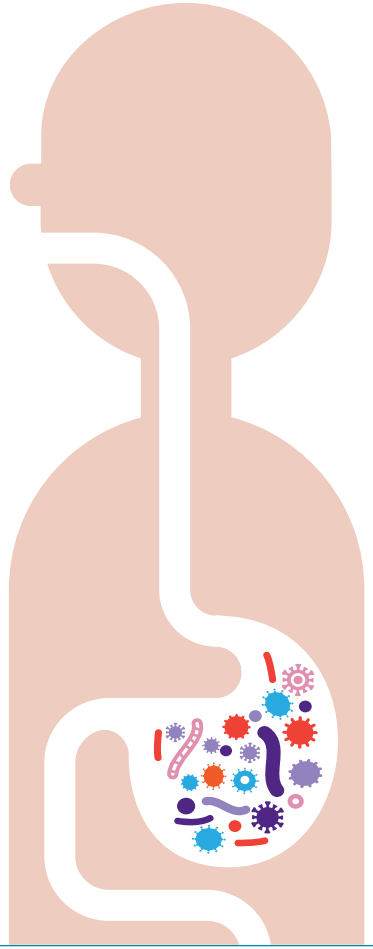
# Known Mechanisms of Action:

**Gut Microbiota**

Changing the gut microbiota can enhance the efficacy of KDT.[1]

A study in mice found that those fed a ketogenic diet had decreased gamma-glutamyltranspeptidase activity, and inhibiting gamma-glutamylation promotes seizure protection. Overall, this study revealed that the gut microbiota modulates host metabolism and seizure susceptibility in mice.



1. Olson C et al The gut microbiota mediates the anti-seizure effects of the ketogenic diet. Cell 2018 May 173 1728-1741. E13

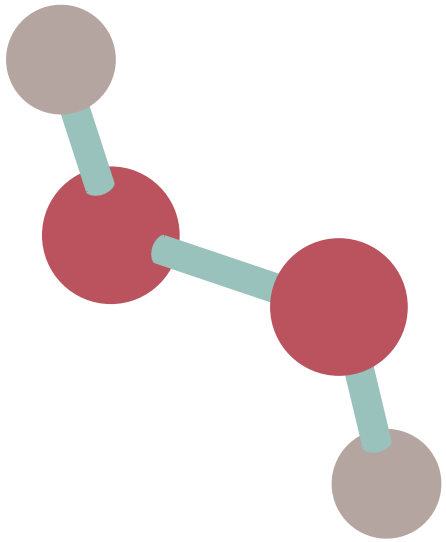
# Known Mechanisms of Action

**Alteration of ion channels**

AMPA receptors – blocked by decanoic acid. This changes the ion channels action potential and reduces excitability[1].

m-TOR pathway, up regulated to promote cell growth, proliferation and survival is suppressed. This reduces synaptic plasticity; inhibiting seizure initiation[2].

Fatty acids / PUFA inhibit voltage gated sodium channels reducing excitability[3].



($H_2O_2$ Hydrogen Peroxide)

Chang P, Augustin K, Boddum K, et al. Seizure control by decanoic acid through direct AMPA receptor inhibition. Brain. 2016;139(Pt 2):431–443. doi:10.1093/brain/awv325
McDaniel SS, Wong M. Therapeutic role of mammalian target of rapamycin (mTOR) inhibition in preventing epileptogenesis. Neurosci Lett. 2011;497(3):231–239. doi:10.1016/j.neulet.2011.02.037
Elinder F, Liin SI. Actions and Mechanisms of Polyunsaturated Fatty Acids on Voltage-Gated Ion Channels. Front Physiol. 2017;8:43. Published 2017 Feb 6. doi:10.3389/fphys.2017.00043

# Possible Side Effects and their Management

# Side Effects

**Keto Flu**

When starting KDT some people may have the following symptoms: nausea[1], fatigue and headache. Although these symptoms usually pass in a few days it is helpful if patients and carers are aware this can occur.

Gradual reduction in carbohydrate when starting the ketogenic diet can help minimise keto flu especially when there is a previously high carbohydrate intake. Also a gradual increase in fat rather than advising the full ketogenic dietary prescription.

1. Neal et al Ketogenic Diet as a treatment of childhood epilepsy; a randomised controlled trial. Lancet Neuro 2008; 7:500-506

# Side Effects

**Constipation[1]**

This is a common side effect of the ketogenic diet due to it being low fibre. It is advisable to explain that this maybe a side effect when commencing a ketogenic diet as constipation can often affect seizure control.

It is recommended that adequate fluids are given, and offering some of the lower carbohydrate food which are higher in fibre eg. ground flax seed, chia.

Resource Optifibre can sometimes help and this can be added to fluids and also given as a flush for children who are PEG fed. A prescription for Paediatric Movicol may also be needed.

1. Neal et al Ketogenic Diet as a treatment of childhood epilepsy; a randomised controlled trial. Lancet Neuro 2008; 7:500-506

# Side Effects

**GI symptoms**

eg. gastro oesophageal reflux can be a side effect of the ketogenic diet, as a high fat diet can cause delayed gastric emptying.[1]

A lower ratio diet can often be helpful, and smaller, more frequent meals. If the child is previously prescribed anti reflux medications then these need to be maximised. If symptoms are severe then advice from a gastroenterologist can be helpful.

1. El-Rashidy OF et al. Modified Atkins diet vs classic ketogenic formula in intractable epilepsy. Acta Neurol Scand. 2013 Dec;128(6):402-8

# Side Effects

**Raised lipids**

It is important to record a baseline level to ensure any lipid abnormalities prior to KDT are picked up.

The lipid profile should be monitored regularly. If the lipids are particularly high then it may help if the saturated fats are replaced by mono-unsaturated and polyunsaturated fats.

A study found that even though total cholesterol and LDL levels increased in adults during the first three months of MAD, within a year cholesterol levels normalised[1].

1. Cervenka MC et al The impact of the modified Atkins diet on lipid profiles in adults with epilepsy. Nutr Neurosci. 2016;19(3):131-7

# Side Effects

**Renal stones**

Renal stones may occur in a small percentage of patients, especially if they are having poor fluid intake. A low carbohydrate diet can have a diuretic effect.

Some ASMs that are carbonic anhydrase inhibitors such as Topiramate[1] may increase the risk of renal stones. A renal ultra sound before starting a ketogenic diet is advisable, and important to encourage adequate fluids.

1. Sampath et al Kidney stones and the ketogenic diet: risk factors and prevention. J Child Neurol 2007; 22:375-378

# Side Effects

**Growth[1]**

Poor growth has been documented for children having a ketogenic diet for a long time. Acidosis is a possible reason for this.

It is important that children following a ketogenic diet are regularly monitored by an experienced dietitian who will monitor their height and weight.

1. Raju KNV et al Efficacy of 4:1 (classic) versus 2.5:1 ketogenic ratio diets in refractory epilepsy in young children: a randomized open labeled study Epilepsy 2011 Sep;96(1-2):96-100

# Side Effects

**Increase risk in fractures**

Chronic acidosis and poor calcium and vitamin D[1] intakes can be factors.

1. Bergqvist AGC et al Progressive bone mineral content loss in children with intractable epilepsy treated with ketogenic diet. AM J Clin Nutr 2008;88:1678-1684

# Module 3
## Practical Considerations for KDT and Supporting Information

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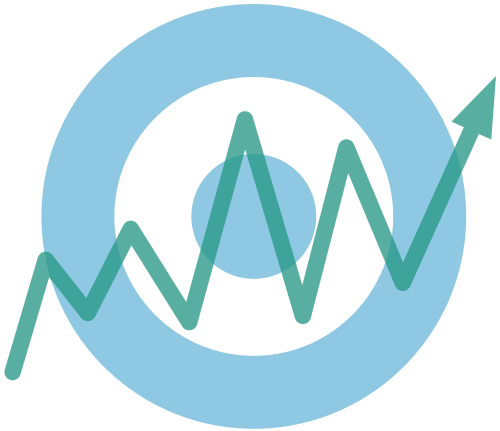
# Monitoring and Quality of Life Measures

# How is KDT Monitored?

- Daily ketone readings.
- Daily seizure diary.
- Evaluating changes in quality of life.
- Regular weight record.



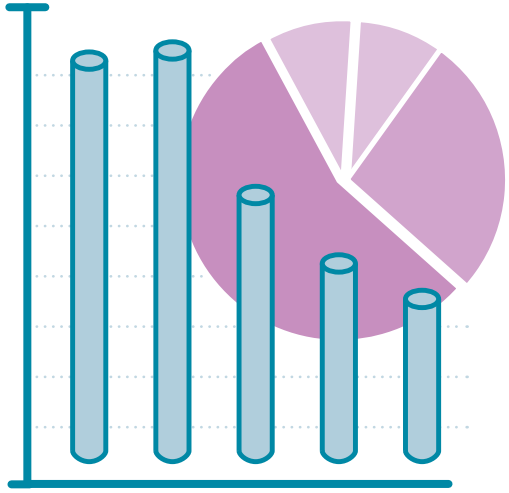
# Ongoing Monitoring

There tend to be more queries and concerns during the first few weeks so the following can all be discussed effectively and therapy adjusted accordingly via regular phone or email contact with the team:

- Ketone monitoring.
- Diet changes.
- Seizure changes.
- Growth (weight rather than height).

Hospital appointments are important to review the following:

- Seizure control.
- Triglyceride and cholesterol profiles.
- Blood tests to check vitamin and mineral status.
- Bone health scans.
- Kidney scans to check for stones.



# How are Ketones Measured?

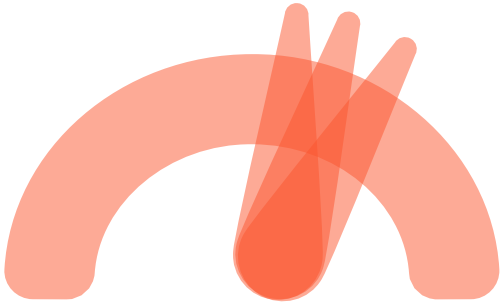
- In blood using a home monitoring device.
- In urine using urine dipsticks.

# Why are Ketones Measured?

- To allow for optimisation of dietary therapy.
- To ensure ketone levels do not become excessive. Symptoms of this occurring are irritability, nausea and vomitting.

# What can Cause Ketone Levels to Fluctuate?

- Not enough fat in the diet.
- Too little or too much energy in the diet.
- Growth spurt.
- Unexpected sources of carbohydrates in the diet e.g. medicines, sweeteners, birthday parties and celebrations at school or within the family.
- The time of the day that ketones are measured e.g. ketones can be lower in the morning compared to the evening.
- Often ketone levels fall prior to and during illness/infection.



# Monitoring Seizures

Encourage a daily record of seizures, which can be classified by type, frequency, duration and severity.

Occasionally an increase in seizure frequency may be observed during the initiation phase of KDT. If the child is unwell with worsening of seizures a clinical review is required.

The seizure and ketone diary should be reviewed regularly by the KDT consultant, dietitian, nurse and family to determine if the diet is being effective.

Approximately half of children who trial the ketogenic diet will experience a fifty percent reduction in seizures.

KDT should be trialled for a least 3 months to gauge efficacy

# Quality of Life

When only a small reduction in seizures is observed, it is important to monitor and evaluate any of the following additional benefits KDT may provide:

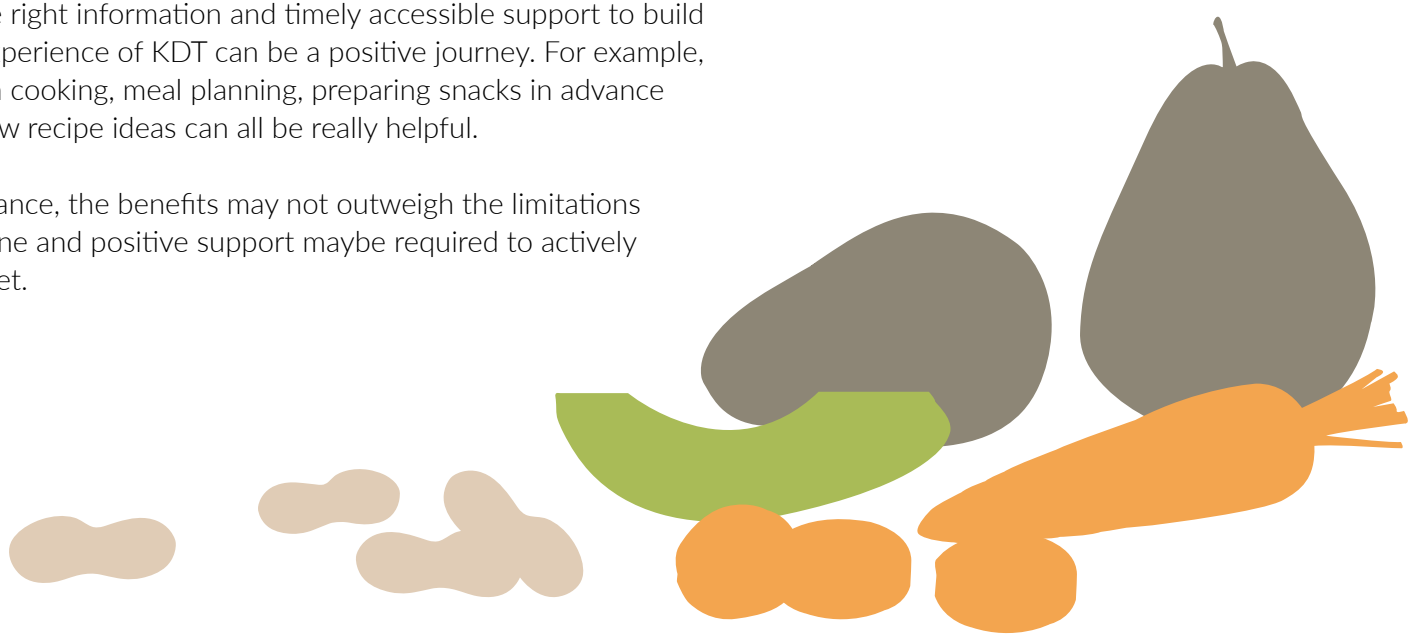
- Reduction in seizure intensity.
- Improved levels of alertness and concentration.
- Improved sleep.
- Improved changes in behaviour or communication.
- Even if there has only been a small reduction in seizures
- If overall QoL has improved then the family may wish to continue KDT with the clinician's consent.

# Overall Evaluation of Efficacy of KDT after 3 Month Trial Period

Following KDT brings a significant lifestyle change and is a steep learning curve. Understandably, many familes and adult patients need considerable support from their keto team, especially during the first few weeks.

However, with the right information and timely accessible support to build confidence, the experience of KDT can be a positive journey. For example, encouraging batch cooking, meal planning, preparing snacks in advance and generating new recipe ideas can all be really helpful.

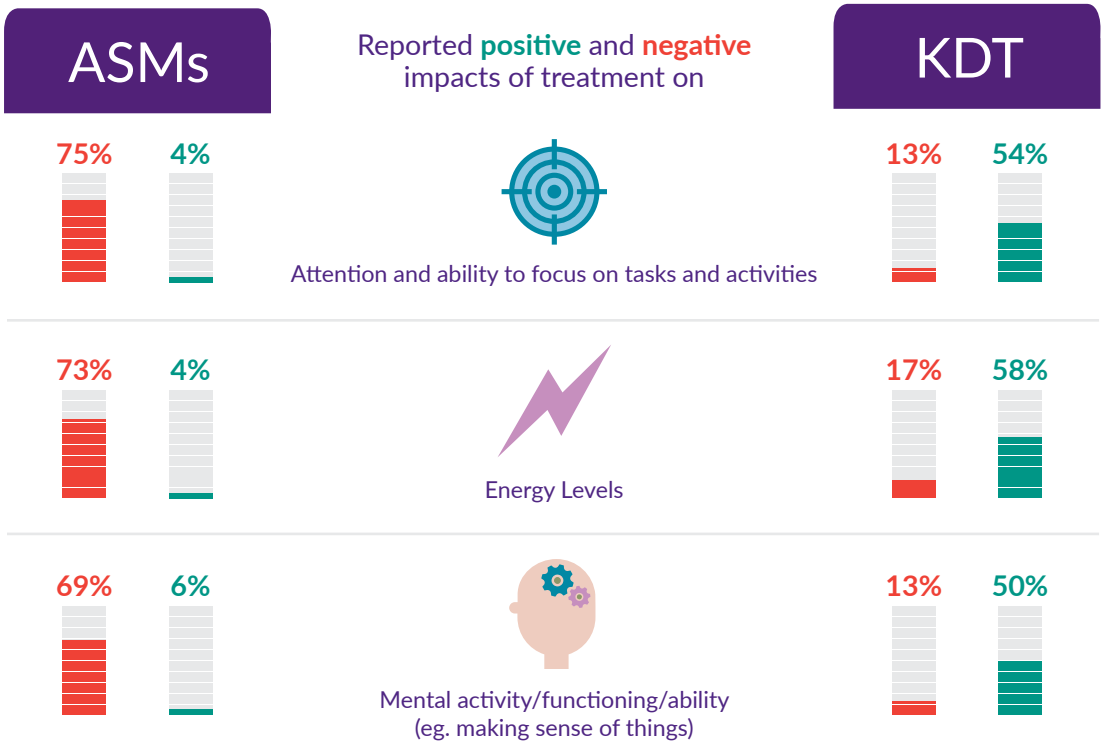
Sometimes on balance, the benefits may not outweigh the limitations of KDT for everyone and positive support maybe required to actively discontinue the diet.



# Positive & Negative Impacts of Treatment - ASMs vs KDT

Two thirds of parents see negative side effects with ASMs.

By contrast, the KDT positively impacts the energy, alertness, communication and focus - all things that contribute to overall quality of life.




1. The Impact of Epilepsy and its Treatment on Quality of Life from a Parental Perspective – VJ Whiteley et al Journal of Developmental Medicine & Child Neurology Volume 61 S1 Jan 2019 p42–43 (99)
2. Coping With Complex Epilepsy Whilst Striving for a Quality of Life for the Whole Family – Facilitating a Parent's Perspective. Neurodigest, Issue 4, Winter 2018 p15–16

# Patient Benefits and Target Setting

# Before Starting KDT

It is recommended to determine what aims the patient and or family want to achieve in commencing KDT. Having aims at the beginning helps everyone to see if outcomes are achieved at review appointments.

**The following are common aims for those trialling the ketogenic diet:**

- To reduce the frequency and severity of seizures and/or to stop seizures.
- To reduce or stop ASMs.
- To reduce the frequency and need for rescue medication.
- To reduce hospital admissions.
- To improve cognitive development.
- To improve school performance and attendance at school.
- To improve sleep.
- Young people and adults may have their own aims for trialling KDT and for example they may hope:
  - » KDT will give them some more independence.
  - » KDT will improve their concentration and so help with their school work.

# Medicines Management

# Do Medicines Contain Carbohydrate?

- Most medicines contain some carbohydrate.
- Liquid medications containing sugar should be avoided.[1]
- Before children start KDT, review their medications and update to the most appropriate formulation.

1. McArtney R et al What is a ketogenic diet and how does it affect the use of medications. Arch Dis Child Educ Pract Ed 2017;102:194–199

# Which Words Mean Carbohydrate?

All ingredients in medications are listed on the patient information leaflet included in the package – words to look out for include:

- Dextrose
- Fructose
- Glucose
- Glycerol
- Lactose
- Maize starch
- Cellulose
- Chitin
- Sucrose
- Mannose
- Galactose


Contact your hospital pharmacist or medicines helpline for further information.

# Which Type of Medicines are Suitable?

- Tablets and capsules are generally better than liquids as they contain fewer carbohydrates.

# What About Children Unable to Swallow?

- For children who cannot swallow and a liquid equivalent is not available, offer a crushable option.
- Teach families how to disperse the medicine in a small amount of water.

# Are Sugar Free Medicines Suitable?

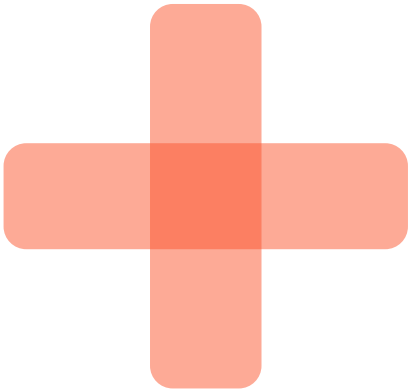
- Some sugar-free liquid medications contain sorbitol, which can affect ketone levels.
- If in doubt, request medicines in tablet form.

# What About Suppositories?

- Suppositories are safe to give as carbohydrate is not absorbed by this route.

# Helpful Hints During Hospital Admission

- Intravenous fluids, often contain glucose (dextrose) so these should be avoided unless the medical team feels they are necessary.
- Please contact the ketogenic diet team if you are unsure.
- Antibiotics should be prescribed in tablets where possible.
- If a tablet form is not available, give the liquid medication as illness management takes priority.
- Change to tablet or low carbohydrate version as soon as possible and monitor ketones twice daily.
- Make sure an alert is added to any electronic prescribing system.



# Support and Considerations for the Wider Family and Carers

# Matthew's Friends

Matthew's Friends charity are global advocates of medical Ketogenic Dietary Therapies, since 2004, working and networking with key professional Ketogenic experts from around the world.

With the main organizational hub in the UK, the charity provides extensive information and support for families, patients and professionals alike, whilst also supporting a clinical service for both children and adults via the Matthew's Friends Clinics UK, for where there is no local service available. They provide in-depth CPD courses for health care professionals by way of an annual KetoCollege training meeting and KetoCollege Online tutorials for those new to Keto and from low-income countries. Matthew's Friends have appropriately placed funding into numerous NHS ketogenic centres and dietetic posts throughout the UK and Ireland, as well as funding for clinical research projects. The charity is also instrumental in organizing the bi-annual Global Symposia on Medical Ketogenic Dietary Therapies and are founding members of the International Ketogenic Neurological Society (INKS).

Family support is key to compliance and success of the diet and to aid this the charity provides numerous free resources; including a comprehensive website and extensive recipe section, as well as YouTube Channels featuring expert educational films and a popular KetoCooking Channel. Matthew's Friends provide a thriving, secure, patient/family support forum accessed via Facebook, a telephone Keto support line, hugely popular live via Zoom virtual 'Cuppa and Chat' sessions for families to be able to join where ever they may live, with guest professionals joining regularly so that families and patients can 'ask the expert' directly. They also offer a bespoke recipe development service for families and dietitians alike and all UK/Ireland patients are entitled to a free Keto starter pack, which contains useful equipment and guidance along with free samples (where appropriate), which help prepare families commence their Ketogenic journey. For those families that are really struggling or having difficulty coming to terms with the situation they are finding themselves in, the charity also funds online 'Mindfulness for Anxiety' courses and again, these have proved extremely useful for families.

For further information about the charity and all their available resources please visit *www.matthewsfriends.org*

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

MATTHEW'S FRIENDSCLINICS
KETOGENIC DIETARY THERAPIES

Matthew's Friends KetoCollege®

# Daisy Garland

The Daisy Garland is a national, registered charity working exclusively for children and young adults with drug-resistant epilepsy. Their drive and determination over the last 17 years has been instrumental in bringing the ketogenic diet to the forefront in the UK.

Working in partnership with 15 NHS trusts, The Daisy Garland has funded 26 ketogenic dietitians giving 1000's of children and young adults the opportunity to access, free of charge, the ketogenic dietary therapy.

Parent support is their top priority and you will find this in abundance in Daisy's Keto Café. Accessed via Facebook, this private ketogenic support group is a safe place for families to exchange recipes, share experiences and talk over worries and concerns.

Their passion for raising ketogenic awareness knows no bounds. Complimentary Daisy Welcome Boxes, sent to every new keto family are packed with keto-friendly food samples and all the essentials needed to start and maintain the diet. They also provide a wide range of free ketogenic information and resources for families and healthcare professionals and their award-winning charity film clearly demonstrates the impact of their vital work within the ketogenic sector.

The Daisy Garland charity helpline and website offer enhanced ketogenic support where quick and easy keto recipes and inspiring cookery demonstration videos can be viewed www.thedaisygarland.org.uk

Nutricia is proud of its long partnership with The Daisy Garland, during this time we have:

- Collaborated with Daisy Garland ketogenic Dietitians to highlight the advancement of the ketogenic dietary therapy
- Helped create ketogenic recipes and cookery demonstrations inspiring and encouraging ketogenic adherence
- Collaborated with The Daisy Garland to measure quality of life for families following the ketogenic diet

The Daisy Garland
*'A journey of hope... for children with epilepsy'*

# Nutricia Product and Service Information

# Ketocal 3:1 Powder

A 3:1 ratio powder with an advanced fat profile.
Suitable as the sole source of nutrition or for supplementary intakes from birth.

- Available in 300g tins.
- Versatile: easy to mix powder.
- Available in unflavoured.
- Can be used in all forms of KDT: Classical, MCT or MKD.
- Contains carnitine to support B-oxidation.
- Carnitine plays an important role in energy metabolism[1] and can be deficient in patients taking certain ASMs.[2]
- Advanced fat profile: reduces the intake of saturated fat for long-term health benefits and includes EFAs and LCPs.

**KetoCal 3:1 Powder** provides a convenient way of administering KDT: as a drink, in meals or as a tube feed[1]


1. Flanagan, J.L, Simmons P.A. et al. (2010) 'Role of Carnitine in Disease', Nutrition & Metabolism, 7 (30), pp 1743-1775
2. Coppollo, G, Epifanio, G. et al. (2006) 'Plasma Free Carnitine in Epilepsy Children, Adolescents and Young Adults Treated with Old and New Antiepileptic Drugs with or without Ketogenic Diet', Brain and Development, 28 (6), pp 358-365

# Ketocal 4:1 Powder

A 4:1 ratio powder with an advanced fat profile. Suitable as the sole source of nutrition for 1 years+ or for supplementary feeding.

**KetoCal 4:1 Powder** provides a convenient way of administering KDT: as a drink, in meals or as a tube feed[1]


- Available in 300g tins.
- Versatile: easy to mix powder.
- Two flavours: unflavoured and vanilla.
- Can be used in all forms of KDT: Classical, MCT or MKD.
- Contains a unique multi fibre blend to help improve bowel health as fibre intake is reduced on a KDT[1] carnitine to support B-oxidation.
- Carnitine plays an important role in energy metabolism[2] and can be deficient in patients taking certain ASMs.[3]
- Advanced fat profile: reduces the intake of saturated fat for long-term health benefits and includes EFAs and LCPs.

1. Bergqvist AG. Long-term monitoring of the ketogenic diet: Do's and Don'ts. Epilepsy Res. 2012;100(3):261–6
2. Flanagan, J.L, Simmons P.A. et al. (2010) 'Role of Carnitine in Disease', Nutrition & Metabolism, 7 (30), pp 1743-1775
3. Coppollo, G, Epifanio, G. et al. (2006) 'Plasma Free Carnitine in Epilepsy Children, Adolescents and Young Adults Treated with Old and New Antiepileptic Drugs with or without Ketogenic Diet', Brain and Development, 28 (6), pp 358-365.'

# Ketocal 4:1 LQ

A 4:1 ratio ready to drink liquid suitable as the sole source of nutrition for 1 years+ or for supplementary feeding.

**KetoCal 4:1 LQ** provides a convenient way of administering KDT: as a drink, in meals or as a tube feed[1]


**Key features:**

- Available in 200ml cartons.
- Two flavours: unflavoured and vanilla.
- Quick and easy: ready to drink liquid.
- Can be used in all forms of KDT: Classical, MCT or MKD.
- Contains a unique multi fibre blend to help improve bowel health as fibre intake is reduced on a KDT.[1]
- Advanced fat profile: reduces the intake of saturated fat for long-term health benefits.

1. Bergqvist AG. Long-term monitoring of the ketogenic diet: Do's and Don'ts. Epilepsy Res. 2012;100(3):261–6

# Introducing the First Ketogenic Diet Formula Specifically Designed for Adolescents and Adults

A 2.5:1 ratio ready to drink liquid suitable as the sole source of nutrition or for supplementary feeding

- Available in 200ml cartons.
- Great vanilla flavour.
- Convenient and easy to use ready to drink liquid.
- Can be used in all forms of KDT: Classical, MCT or MKD.
- Contains contains a unique, multifibre blend to help meet daily fibre needs and support gut health ref.[1–7]
- Balanced fatty acid profile.
- Suitable for complete nutritional support from 8 years+.

**KetoCal 2.5:1 LQ** provides a convenient way of administering KDT: as a drink, in meals or as a tube feed


1. Green, C.J. (2001) 'Fibre in Enteral Nutrition', Clinical Nutrition, 20(1), pp. 23–39.
2. Trier, E. et al. (1999) 'Effects of a Multifibre Supplemented Paediatric Enteral Feed on Gastrointestinal Function', Journal of Paediatric Gastroenterology and Nutrition, 28(5), pp. 595.
3. Evans, S. et al. (2009) Journal of Human Nutrition and Dietetics, 22, pp. 414–421.
4. Hofman, Z. et al. (2001) Clinical Nutrition, 20 (S3) pp. 63.
5. Daly, A. et al. (2004) Journal of Human Nutrition and Dietetics, 17, pp. 365–70.
6. Guimber, D. et al. (2007) Journal of Paediatric Gastroenterology Nutrition, 44 pp. 201.
7. Grogan, J. et al. (2006) Journal of Human Nutrition and Dietetics, 19 pp. 458–477.

# MCT Oil

A 100% MCT fat emulsion that can be used as part of a modular feed.

**Key features:**

- Suitable for all ages.
- 500ml resealable plastic bottles.
- Suitable for cooking.
- MCT Oil can be mixed with liquids or used as a cooking ingredient.
- MCT Oil should be introduced slowly into the diet as rapid absorption of large intakes may cause vomiting or diarrhoea.

# Liquigen

A 50% MCT fat emulsion that can be used as part of a modular feed.

**Key features:**

- Suitable for all ages.
- 50% MCT fat emulsion.
- 250ml resealable plastic bottles.
- Can be easily flavoured.
- Liquigen can be mixed with liquids, used as a milk substitute or used as a cooking ingredient.
- Liquigen should be introduced slowly into the diet as rapid absorption of large intakes may cause vomiting or diarrhoea.

# Phlexy-Vits Tablets / Sachets

- An unflavoured powder that provides vitamins, minerals and trace elements. Comes in 7g dose sachets or tablets (180 per tub).
- One sachet can help meet the nutritional needs of patients aged eleven and older. Powder can be measured to meet the needs of younger children.
- Flexible formats for added convenience and adaptable to individual needs.
- 0.5g carbohydrate per 100g - low in carbohydrate for patients following a ketogenic diet.

# Calogen

A ready to feed, high energy, neutral flavoured LCT fat emulsion.

**Key features:**

- Suitable for all ages.
- 50% LCT fat emulsion.
- Available in 200ml and 500ml bottles.
- Ready to feed liquid.
- 0.1g carbohydrate per 100ml.
- Calogen can be mixed with liquids, used as a milk substitute, used as a cooking ingredient, or added to modular feeds for tube feeding.


Calogen is a Food for Special Medical Purposes for the dietary management of conditions requiring a high energy intake and must be used under medical supervision.

# Protifar

A versatile easy to mix powder, low in carbohydrates, high in protein.

**Key features:**

- Suitable for 3 years and =.
- Available in 225g tins.
- Versatile: easy to mix powder, low in carbohydrate.
- Available in neutral flavour.
- Protifar can be used as part of a modular feed and is adaptable for use with tube fed patients.


Protifar is a Food for Special Medical Purposes for the dietary management of hypoproteinaemia and must be used under medical supervision.

# Maxijul

A versatile powdered carbohydrate supplement.

**Key features:**

- Suitable for all ages.
- Available in a 200g can or 132g sachets.
- Versatile: easy to mix powder.
- Unflavoured.
- Can be used in multiple ways to help fine tune the ketogenic diet: add to liquids, foods or modular feeds.
- Used to manage high ketone levels and episodes of low blood sugar.


Maxijul is a Food for Special Medical Purposes for the dietary management of disease related malnutrition or any condition where additional energy from a carbohydrate source is required and must be used under medical supervision.

# MyKetoPlanner

The new online tool helping to provide flexibility and variety for the ketogenic community. Available at MyKetoPlanner.co.uk or MyKetoPlanner.ie.

- Create recipes and daily meal plans for patients.
- Search and edit recipes shared by other families or dietitians.
- Mobile and tablet friendly sites to check on the go.