

KETOGENIC DIETS FOR GLUT1 DEFICIENCY

GLUCOSE TRANSPORTER TYPE 1 DEFICIENCY SYNDROME

TIPS FROM:
Victoria Whiteley, RD
Royal Manchester Children's Hospital
Glut1 Deficiency UK Medical Advisory Board

Medically supervised ketogenic diets are the current standard of care treatment for Glut1 Deficiency, but there are special considerations for their use in this disease.

Managing the triggers of symptoms:

Despite the ketogenic diet being the recommended management option, it does not provide 100% symptom management. The symptoms and triggers of GLUT1 Deficiency change with age. Most often symptoms occur before meals, with tiredness, on waking and are exacerbated by fasting or exercise. Symptoms can also be triggered by drops or fluctuations in ketones levels. There are also other triggers which are harder to manage including emotional stress (anxiety, upset, excitement, stress), fever/illness, fatigue, environmental changes (temperature) and routine changes.

Triggers:	Tips:
Fasting	<ul style="list-style-type: none">• Ensure regular pattern of meals and snacks
Exercise	<ul style="list-style-type: none">• Include exercise as part of the daily routine• Add in additional snacks for high intensity exercise (swimming)
Fatigue/sleep deprivation	<ul style="list-style-type: none">• Ensure activities are well paced• Employ good sleep hygiene
Environmental temperature	<ul style="list-style-type: none">• Consider holiday destinations (choose air conditioning)• Plan activities around weather• Drink well and maintain good ketosis• May need to adjust to small frequent keto meals/snacks if appetite impacted
Emotional stress	<ul style="list-style-type: none">• Extra snacks• Clinical psychology support around managing anxiety/excitement/stress• Support from school in managing change• Holistic approach to behavioral management

Managing the obstacles of the ketogenic diet:

Maintaining good ketosis at all times can be difficult. Regular dietary review is required as needs change with age and growth.

Obstacles:	Tips:
Low morning ketones	<ul style="list-style-type: none"> • Additional fats at bed time
Constipation (can lower ketones)	<ul style="list-style-type: none"> • Increase fibre (flaxseed/avocado/rhubarb/berries/nuts/psyllium husk), fluid, toileting routine, probiotics, consider stool softeners
Reflux or vomiting	<ul style="list-style-type: none"> • Smaller more frequent meals and snacks • Change types of fat, reducing MCT
Growth or weight changes	<ul style="list-style-type: none"> • Regular dietetic review. If calories are too high or too low ketones can be impacted.
Medications	<ul style="list-style-type: none"> • Sugar free liquid meds may contain sugar alcohols (e.g. Sorbitol) which can impact on ketosis
Teething	<ul style="list-style-type: none"> • Pain relief and soothing gums
Illness and hospital admissions	<ul style="list-style-type: none"> • Agree a plan that works for your child, may include a shake to replace meals • Clear information from the team about inpatient management
Flexible/modified plans	<ul style="list-style-type: none"> • Can be great to improve compliance long term, but be careful of protein intake as too much can reduce ketones
Food refusal	<ul style="list-style-type: none"> • It is a normal part of development and childhood • Similar meal types • Food rules are the same for the whole family • Reward positives
School	<ul style="list-style-type: none"> • Education and training of school staff • Plan ahead for meals, snacks and activities
Parties, celebrations and social events	<ul style="list-style-type: none"> • Plan ahead • Take meals, snacks, treats • New recipes online from places like Matthews Friends and Charlie Foundation
Holiday and travel	<ul style="list-style-type: none"> • Plan ahead • Investigate what is available where you are going and what you will need to take and how
Hormones	<ul style="list-style-type: none"> • For girls, an extra snack during ovulation and menstruation • For boys, may need regular review to meet rapid growth needs

Compliance:

Ketogenic diets are hard. The relationship between knowledge and adherence is not as strong or direct as is assumed in many theories of health behaviour change. Because we know, doesn't mean we do. There is more non-adherence in adolescence (which is part of normal development) and less strict diets, like modified plans may be better tolerated long term.

Ways to improve compliance:

- Praise honesty
- Avoid blame and acknowledge how difficult adherence/ compliance is
- Explore understanding
- Explore motivations
- Encourage questions
- Provide advice, info, and support in accessible formats
- Identify worries, concerns, and barriers
- Praise achievements and build on successes
- Peer support is really important, its easier to accept info/advice from peers
- Networking gives you a feeling you belong, and others understand
- Cookery sessions/family days/youth meetings/sibling support

Children that do well:	Parents that do well:
Optimism for condition and life in general	Have a confidante to talk to
Young people who perceive themselves to be in control of situations generally	Remind themselves they are doing the best job they can (self-compassion)
Young people who are good at self-monitoring and have problem-solving strategies	Stay calm, have stable routines but can be flexible

Care Coordination:

Working with a multi-professional team allows for comprehensive and co-ordinated care, improving efficiency of care and reducing the burden on the family.

- Doctor (consultant in neurology or metabolics)
- Primary care physician
- Ketogenic Dietitian (ideally with GLUT1 Deficiency experience)
- Physiotherapy to support tone and movement
- Speech and language therapist
- Occupational therapist to support fine and gross motor skills
- Clinical psychologist to assess learning and psychological support needs
- Education team to ensure appropriate education assessment and support