

## KETOGENIC DIET FOR GLUT-1 DS

### SUMMARY OF 2015 FAMILY CONFERENCE PRESENTATION

#### OVERVIEW

This is a summary of information provided at the Glut-1DS Family Conference in 2015. Newer information may precede the guidelines in this document.

Glucose normally fuels the brain. In the ketogenic diet, ketones rather than glucose are the main source of fuel for the brain. Ketogenic diet therapies should be supported by a knowledgeable nutritionist and supervised by a physician. Laboratory monitoring is advised to monitor health status and prevent adverse effects.

Ketogenic diet therapies include the following:

- $\beta$ -OHB monitoring
- Diet
- Fluids
- Supplements
- Laboratory monitoring
- Super Foods

#### $\beta$ -OHB MONITORING

$\beta$ -OHB stands for betahydroxybutyrate which you will monitor for in the blood. Purchase or obtain a “Precision Xtra” meter at a pharmacy or online ([abbott.com](http://abbott.com)); the cost is approximately \$75. The meter comes with a few strips. Additional strips are approximately \$3–6 each. There is potential coverage through insurance (with letter of medical necessity). Urine ketone testing is not necessary if using the monitor.

When to check  $\beta$ -OHB Levels.

- During the initiation of ketogenic diet therapy check daily at the same time.
  - Levels are typically lowest after sleeping and highest after meals and activity.
- When levels are stable, check once a week.
  - Check again if the individual has an increase in seizures, is more ataxic, or is less focused.
- Goal  $\beta$ -OHB Levels:
  - Children 4–5mmol/L
  - Adults < 4mmol/L

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#### DIET

Ketogenic diets are high in fat, low in carbohydrate and moderate in protein. The Classic Ketogenic diet is designed in a ratio of fat to non-fat (carbohydrate and protein) of 4:1 and 3:1 and foods are weighed in grams. These ratios are advised for infants and children until puberty. Less restrictive ratios of 2:1 and 1:1 can be managed without weighing foods and are easier for older children and adults.

- Adjust the ratio of fat to carbohydrate + protein until  $\beta$ -OHB range is 4-5mmol/L.
  - Infants may not reach 4-5mmol/L. Glucose should also be checked in infants.
  - Children can become ketotic with lower fat to carb + protein ratios such as 3:1.
  - Work with nutritionist to adjust ratio to fit the patient's protein requirements.
  - Carnitine may be a helpful prescription supplement to achieve higher ketones.
  - Medium chain triglycerides (MCT) may help to boost ketosis.
  
- Adjust calories to maximize ketosis:
  - Avoid obesity by consuming sufficient but not excessive calories. Ketogenic snacks may be used to flex calories up for active children.
  - Calories should be established to ensure there is growth, and that height and weight are proportional. Children grow at a predictive rate from birth through puberty.
  - Weight and height should be measured regularly:
    - Infants under 6 months – daily.
    - Infants older than 6 months – weekly.
    - Toddlers – every 2 weeks.
    - Older children – every month.

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## FLUIDS

Ketogenic diets cause the kidneys to release fluid more efficiently which can cause dehydration. It is important to know how much fluid is needed to prevent dehydration. Dehydration that occurs frequently can lead to feeling sluggish, cause headache, constipation and even kidney stones.

Caffeine should not be consumed by individuals with Glut1-DS. Caffeine interferes with glucose transport into the brain.

### Fluid Requirements

The chart shows an average amount of fluid needed for each age group. The specific amount needs to be individualized. People living in warm climates need more water than others. More fluid may be needed during illness with fever, vomiting or diarrhea. The best fluid is water.

<b>Infants</b>	Up to 1000mL (4 cups)
<b>Young children</b>	1000-1500mL (4-6 cups)
<b>Older children</b>	1500-2000mL (6-8 cups)
<b>Adults</b>	2000-2500mL (8-10 cups)

### Electrolyte Replacement Fluid for Sick-Days or During Fasting

<b>Name and Manufacturer</b>	<b>Electrolytes</b>	<b>Measure</b>	<b>Minerals</b>	<b>Directions</b>
Morton Lite Salt	Potassium (K) Chloride (Cl)	1/8 + 1/4 teaspoons	525mg K 1133mg Cl	Dissolve salts into water. Use within 1 day. Does not need to be refrigerated.
Baking Soda	Bicarbonate (CO <sub>3</sub> ) Sodium (Na)	1/2 teaspoon	1670mg CO <sub>3</sub> 1050mg Na	
Water - room temperature, purified water (not distilled)		1 liter (4 c.)		

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## NUTRITIONAL SUPPLEMENTS

Ketogenic diets are low in many vitamins and minerals and for this reason supplementation is necessary. The supplements should have zero or minimal carbohydrate content. Two products that contain a wide array of vitamins, minerals and trace minerals are listed below. Another option is to use a combination of several different products. Ask your nutritionist to assist you with selecting the best options. Alpha lipoic acid: Although alpha-lipoic acid had been recommended based on preliminary laboratory studies, newer evidence shows that alpha-lipoic acid supplementation may assist with muscle energy but not with brain energy.

### Full Spectrum - Low Carb Vitamin with Mineral Supplements

Name and Manufacturer	Form	Carbohydrate Content	Features
FruitiVits - Vitaflo	Packets of powder. Dose is determined by age.	300mg per 6gm packet	29 micronutrients plus fiber
NanoVM t/f - Solace Nutrition	Powder with scoop. Dose is determined by age.	20mg per 5.4gm	25 micronutrients

### Supplements Which may be Helpful for Glut-1DS

Name and Manufacturer	Form	Carbohydrate Content	Notes
<b>Vitamin D3</b>	Liquid or capsules	Various brands - without added carb.	Important hormone for multiple biological functions including mental health and bone health.
<b>Omega-3 Fatty Acid</b>	Liquid or capsules	Various brands without added carb	Important for brain function. May be obtained from food sources i.e. eggs, avocados, olive oil, seed and nut oils.
<b>Carnitine</b> Sigma Tao or Generic brand. Prescription required.	330mg tablet	0	Aids in using fat at the cellular level and boosts ketones.
	Sugar-free solution	0	
<b>MCT oil</b> Various brands	Liquid oil	0	Saturated fat source that is easily used by the brain; boosts ketosis. Has a laxative effect in large doses.
<b>MCT oil emulsion</b> 1. Betaquik (Vitaflo) 2. Liquigen (Nutricia)	Oil and water mixture	0	Saturated fat source that is easily used by the brain; boosts ketosis. May be better tolerated than MCT oil. Has a laxative effect in large doses.
<b>Digestive aids</b>	1. Pancreatic enzyme	Capsules	Check with nutritionist
	2. Lecithin emulsifier: Sunflower or Soy	Various forms including liquid and powder	Varies

## **LABORATORY MONITORING**

Laboratory monitoring is recommended to ensure health and prevent potential deficiencies. The following laboratory studies should be drawn prior to starting the ketogenic diet and at 1, 3 and 6 months then every 6 months.

- Complete blood count
- Lipid profile
- Liver/hepatic profile
- Metabolic panel
- Electrolytes
- Calcium
- Magnesium
- Phosphorus
- Selenium
- Carnitine profile

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## SUPER FOODS FOR KETOGENIC DIETS

Optimize ketogenic diets with nutrient-dense foods

Super Food	Unique Content	Tips
<b>Asparagus</b>	Chromium enhances glucose metabolism.	Steam for best flavor or chop raw into salad. Frozen asparagus are often fresher than store-bought fresh.
<b>Arugula</b>	Prevents cholesterol adherence to arteries, has detoxifying and anti-inflammatory benefits.	Baby greens are less bitter than mature. Serve raw, sautéed or blend into smoothies.
<b>Avocado</b>	Magnesium & fiber (both prevent constipation), more potassium (K+) than banana (K+ is an acid buffer), omega-3 fats, anti-inflammatory; sterols – inhibits cholesterol absorption; glutathione – powerful antioxidant.	The pear shaped, Mediterranean variety is higher in fat than oval type. An avocado is ripe when its skin is a brown dark-green color and there is a little “give” when you gently press your thumb into it. Place unripe avocados on the counter to ripen for a couple of days, separating them to allow them space to release carbon dioxide.
<b>Blackberries</b>	Highest antioxidant content of all fruit. High in fiber.	Buy fresh when they are on sale and freeze in quantities.
<b>BRASSICA VEGETABLES:</b> Broccoli Brussels Sprouts Cabbage, Cauliflower, Kale	Sulforaphane blocks a destructive enzyme that damages cartilage; toxic to cancer stem cells. Indols repair DNA in cells.	Boiling decreases the level of sulforaphane; however, steaming, microwaving, or stir frying does not result in significant loss. Sauerkraut (cabbage) has additional nutritional benefits and is low carb.
<b>Celery</b>	Flavonoids; anti-inflammatory, anti-oxidant, immune system enhancing, cholesterol lowering.	Eat raw or blend into smoothies.
<b>Celeriac</b> (celery root)	Antioxidants and phosphorus (acid buffer)	Use raw in salads or coleslaw, or cooked in soup, or as faux mashed potatoes.
<b>Garlic</b>	Potent antiviral, antibacterial and cholesterol lowering.	Dehydrated minced garlic is economical and easy to rehydrate.
<b>Green tea</b>	Phytochemical epigallocatechingallate improves blood flow and lowers cholesterol.	Contains 45mg of caffeine per cup and negligible carbohydrate. Steep green tea in boiling water no more than 2 minutes to prevent bitterness.
<b>Olive oil and olives</b>	Phytochemicals are anticancerous; Oleuropein is a potent free radical scavenger protecting heart tissue.	Avoid olives cured in lye. Seed pure olive oils, not one that is cut with oil (less expensive brands).
<b>Radishes</b>	Anthocyanins have anti-inflammatory and anti-cancerous properties.	Slice thin for salads. Chop and sauté with onions and cauliflower for faux hash browns. Black radishes are more peppery in flavor.
<b>Spinach</b>	Flavonoids and antioxidants, vitamins A, B2, C & K, magnesium, manganese, folate, iron, calcium & potassium.	Eating raw is the best method to preserve nutritional value. Steaming or quick sautéing are second best.
<b>SPROUTS:</b> Alfalfa, Broccoli Fenugreek LENTILS Pea, Radish, Mung	Rich source of enzymes that combine with vitamins and minerals in essential metabolic pathways. Lysine (branched-chain amino acid) helpful for ketosis. Essential fats.	Grow sprouts inexpensively at home in 5-7 days. Several commercial tray designs make it simple and affordable. Serve over salad or mix into smoothie or stir-fry.
<b>Sunflower seeds</b>	High in poly-phenols and Vitamin E (antioxidants), B-complex vitamins especially folic acid and niacin (enhances brain calming GABA). Also high in choline which is a precursor for neurotransmitters.	Sunflower lecithin syrup is a great fat emulsifier for blended keto beverages and tube-feeding formulas. The high choline content also aids in fat digestion.

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