Ketogenic Diet Basics for the Non-Ketogenic Dietitian

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About the Ambassador
Zahava Turner

Zahava Turner is an Assistant Professor of Pediatrics and Neurology, Board Certified in pediatric nutrition working at the Johns Hopkins Hospital in Baltimore, Maryland since 2005. Prior to that, she worked at Schneider’s Children’s Hospital in New York. Zahava specializes in using the Ketogenic diet for infants and children with epilepsy and has spoken both nationally and internationally at several conferences on the ketogenic and Modified Atkins diet for epilepsy. She is a co-author of 19 publications and the widely-referenced book Ketogenic Diets 6th edition.
Topics

• History of the ketogenic diet
• Overview of the basics
• How to calculate the ketogenic diet
• Helpful tips for monitoring
THE KETOGENIC DIET IN THE TREATMENT OF EPILEPSY*
A PRELIMINARY REPORT
M. O. PETERS, M.D.
MINNEAPOLIS, MINN.

~400 BC 0 1800s 1921 1938 1994 2006+
“No thanks, I’m on a low carb diet.”
Something can be done for the child with EPILEPSY...

Five things give the doctor a warmer glow than the increased hope which he can now offer to the child with epilepsy. Medical science has made a deep, searching, and sympathetic study of this disease... and hope that could hardly seem a new hope but a new outlook for many victims of this condition.

For example, important advances have been made in diagnosing epilepsy in both children and adults... recently by the development and use of the electroencephalograph. With this instrument, the doctor can chart the electrical activity of the brain. This valuable information may point the way to the type of treatment that will bring the best results.

Equally valuable are the anticonvulsant drugs which have been effective in 80 to 90 per cent of patients treated. One of these drugs, diphenylhydantoin, was introduced by Dr. James W. Parker as a result of research many years ago. Since then, many other drugs have been developed for the treatment of epilepsy. As a result of continuing research, other drugs already developed are being re-evaluated for possible use in the treatment of epilepsy. Only the doctor can determine whether each drug will prove beneficial in any particular case, and how they should be used.

Medication alone, however, is not enough. The epileptic child needs special treatment. Understanding, cooperation of the family, school, and others with whom he associates. From this cooperation and guidance—most important—may come the greatest help of all. Medication is necessary and important, but it is not enough. It is the cooperation of the family and others who see the child in daily life that makes it possible for him to live a normal and useful life.

If you know a family in which there is an epileptic child, you can give them the reassuring message that "Something can be done for the child with epilepsy."
November 17 1993

“After the ketogenic diet was started, Charlie did well and began to show a decrease in the amount of seizures he was having. By the time of discharge, the patient had had no seizures for several days.”
Ketogenic Diet Studies Published
SPECIAL REPORT

Optimal clinical management of children receiving the ketogenic diet: Recommendations of the International Ketogenic Diet Study Group

Table 2. Published Efficacy Studies of the Ketogenic Diet, Retrospective and Prospective, with 20 or More Patients, 1998–2008

<table>
<thead>
<tr>
<th>Reference</th>
<th>Study Type</th>
<th>Patients, no.</th>
<th>Age Range, yr</th>
<th>&gt;50% Reduction</th>
<th>&gt;90% Reduction</th>
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<tbody>
<tr>
<td>Vining et al.</td>
<td>Prosp.</td>
<td>51</td>
<td>1–9</td>
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<td>Freeman et al.</td>
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<td>Hassan et al.</td>
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<td>52</td>
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<td>Kankirawatana et al.</td>
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<td>Retros.</td>
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<td>Vaisleib et al.</td>
<td>Retros.</td>
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<td>Kang et al.</td>
<td>Retros.</td>
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<td>Prosp.</td>
<td>20</td>
<td>1–10</td>
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<tr>
<td>Total</td>
<td></td>
<td>1,335</td>
<td>0.3–24</td>
<td>56</td>
<td>24</td>
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</tbody>
</table>

Prosp. = prospective; Retros. = retrospective.

*At 3 months.
BRIEF COMMUNICATION

A blinded, crossover study of the efficacy of the ketogenic diet


The ketogenic diet for the treatment of childhood epilepsy: a randomised controlled trial

Elizabeth G Neal, Hannah Cheffie, Ruby H Schwartz, Margaret S Lawson, Nicole Edwards, Geogiana Fitzsimmons, Andrea Whitney, J Helen Cross

Hopkins Double-Blinded Study

- 12-day study period
  - Start of the diet
  - Children with LGS

- Trend towards saccharin superiority in clinical seizures \((p=0.07)\)
  - Median -34 seizures/day over 12 days \((p=0.003)\)

- Probably an inadequate placebo state due to fasting twice

Review article

Ketogenic diet guidelines for infants with refractory epilepsy

Elles van der Louw a,*, Dorine van den Hurk b, Elizabeth Neal c, Bärbel Leiendecker d, Georgiana Fitzsimmon e, Laura Dority f, Lindsey Thompson g, Maddelena Marchió h, Magdalena Dudzińska i, Anastasia Dressler j, Joerg Klepper k, Stéphane Auvin l, J. Helen Cross m
Topics

• History of the ketogenic diet
• Overview of the basics
• How to calculate the ketogenic diet
• Helpful tips for monitoring
Keto Basics

• High fat, adequate protein and low carbohydrate
• Thought to mimic the metabolic state of fasting
  – By producing ketones
• Works on the mitochondrial level
Comparison

Ketogenic Diet 4:1

- 90% Fat
- 8% Protein
- 2% Carbohydrate

American Diet

- 60% Carbohydrate
- 30% Protein
- 10% Fat
Who gets placed on the diet?

- Patients who fail the traditional anti-convulsant therapy or are poor candidates for epilepsy surgery
- Can be used by all ages
- 3 months
- Average time 1-2 years

<table>
<thead>
<tr>
<th>Absolute</th>
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<tbody>
<tr>
<td>Carnitine deficiency (primary)</td>
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<tr>
<td>Carnitine palmitoyltransferase (CPT) I or II deficiency</td>
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<tr>
<td>Carnitine translocase deficiency</td>
</tr>
<tr>
<td>β-oxidation defects</td>
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<tr>
<td>Medium-chain acyl dehydrogenase deficiency (MCAD)</td>
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<tr>
<td>Long-chain acyl dehydrogenase deficiency (LCAD)</td>
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<tr>
<td>Short-chain acyl dehydrogenase deficiency (SCAD)</td>
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<tr>
<td>Long-chain 3-hydroxyacyl-CoA deficiency</td>
</tr>
<tr>
<td>Medium-chain 3-hydroxyacyl-CoA deficiency</td>
</tr>
<tr>
<td>Pyruvate carboxylase deficiency</td>
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<td>Porphyria</td>
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</table>

<table>
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<tr>
<th>Relative</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inability to maintain adequate nutrition</td>
</tr>
<tr>
<td>Surgical focus identified by neuroimaging and video EEG monitoring</td>
</tr>
<tr>
<td>Parent or caregiver noncompliance</td>
</tr>
</tbody>
</table>
Calories, Protein & Fluid

• No calorie restriction
  – Use clinical judgment when estimating nutrition needs
• Current intake
  – GOAL: Meet adequate calories for growth
• DRI for protein
  – Adjust calories or ratio to meet protein needs
• 100% fluid maintenance
Ratio

• Ratio: Grams of fat: protein and carbohydrate combined
  – Example: 4:1 ratio is 4 grams of fat to 1 gram of protein and carbohydrate combined

• Typical ratio and uses
  – 3:1 Infants, teenagers & compromised patients
  – 4:1 > 2yo

• Higher the ratio the lower the amount of allowed protein and carbohydrates
Meal Plan

Basic Structure
• Heavy whipping cream
• Butter/ mayonnaise/ oil
• Protein
• Fruit or vegetable

Typical menu
• 40g 36% heavy cream
• 24g Chicken Breast
• 11g broccoli
• 21g fat
• 12g lettuce
Products

Formulas that may be utilized for the ketogenic diet include:

• **KetoCal® (Nutricia®)**
  - KetoCal® 3:1 Powder (Unflavored)
  - KetoCal® 4:1 Powder (Vanilla Flavored)
  - KetoCal® 4:1 LQ Liquid (Flavored and Unflavored)

• **RCF® (Abbott®) – Ross Carbohydrate Free Formula - soy based carbohydrate free formula**
  - Used in milk protein allergy
  - Used when carbohydrates must be very limited due to low caloric needs

• **KetoVolve™ (Nutr-e-volution)**
  - Bland flavored powder

• **KetoVie™ 4:1 (CamBrooke Therapeutics™)**
  - Available in chocolate and vanilla flavors
  - Ready-to-feed liquid

The labels for KetoCal®, KetoVolve™ and KetoVie™ indicate these products are intended for use with children ages from 1 year plus.
Modular Products
A variety of modular products may need to be added to ensure nutrient needs are met and ketogenic ratios are correct.

**Lipid**
- Microlipid® (Nestle®) – safflower oil emulsion at 4.5 kcal/mL
- MCT Oil® (Nestle®) – fractionated coconut oil at 7.7 kcal/mL
- Liquigen® (Nutricia®) – MCT emulsion at 4.5 kcal/mL
- Betaquik™ (Vitaflo®) – MCT emulsion at 1.89 kcal/mL
- Carbzero™ (Vitaflo®) – LCT emulsion at 1.8 kcal/mL
- Retail Oils (Olive oil, coconut oil) – variable caloric density

**Carbohydrate**
- SolCarb powder (Solace®) – carbohydrate powder – maltodextrin
  - 3.75 kcal/g
- Polycal™ powder (Nutricia®) – carbohydrate powder – maltodextrin
  – 3.84 kcal/g

**Protein**
- Beneprotein® (Nestle®) – whey protein powder – 6 gm protein in 7 gm powder
- Complete Amino Acid Mix (Nutricia®) – 100% amino acid powder – 8.2 g protein in 10 g powder
Supplementation

• Diet deficient in:
  – B Vitamins
  – Vitamin C
  – Calcium, zinc, phosphorous
  – Fiber
  – Trace minerals
  – Carnitine, selenium

• Supplement with carbohydrate free multivitamin and mineral supplement

Table 4. Supplementation recommended for children receiving the KD

Universal recommendations
  Multivitamin with minerals (and trace minerals)
  Calcium with vitamin D

Optional extra supplementation
  Oral citrates (Polycitra K)
  Laxatives: Miralax, mineral oil, glycerin suppository
  Additional selenium, magnesium, zinc, phosphorus, vitamin D
  Carnitine (Carnitor)
  MCT oil or coconut oil (source of MCT) *
  Salt (sodium to add to modular formulas if used for greater than age 1 year)

All supplements listed should be provided as carbohydrate-free preparations whenever possible.

Estimating Calories Protein & Fluid

Calories
• Dietary Reference Intake (DRI) + Activity Factor (AF)
  – Can use WHO, Schofield...
• 3 day food record
  – Important to compare estimated needs with food record and growth history
• No calorie restriction
• Use clinical judgment regarding patients with obesity and failure to thrive

Protein & Fluid

Protein
• RDA

Fluid
• Maintenance fluid calculation
• No fluid restrictions
• “Fluid Requirement”
  – Keep fluid level the same every day
### Calculating Dietary Units

**Food Science:**
- 1 g fat = 9 kilocalories
- 1g protein and 1 g carbohydrate = 4 kilocalories

<table>
<thead>
<tr>
<th>Ratio</th>
<th>Calories per Dietary Unit</th>
<th>Example</th>
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</thead>
<tbody>
<tr>
<td>2:1</td>
<td>22</td>
<td>((2 \times 9) + (1 \times 4) = 22)</td>
</tr>
<tr>
<td>3:1</td>
<td>31</td>
<td>((3 \times 9) + (1 \times 4) = 31)</td>
</tr>
<tr>
<td>4:1</td>
<td>40</td>
<td>((4 \times 9) + (1 \times 4) = 40)</td>
</tr>
<tr>
<td>5:1</td>
<td>49</td>
<td>((5 \times 9) + (1 \times 4) = 49)</td>
</tr>
</tbody>
</table>
Diet Calculation

• By dividing the dietary units of a given ratio into the determined total calories (age x calories per kg)

• The total grams of protein, carbohydrate, and fat in a given ketogenic diet can be determined.
Example of Calculation

• 5 year old female
  – Wt: 17.7kg (45%)
  – Ht 108 cm (50\textsuperscript{th}%)
  – BMI 50\textsuperscript{th}%

• Based on 3 day food record and EER for healthy children = 85-90kcal/kg
  – 1500 calories per day

• Team determined patient will go on 4:1
Diet Calculation

• Divide 40 into total calories
  – 1500/40 = 37.5 Dietary Units

• Multiply Dietary Units by fat grams in ratio
  – 37.5 x 4 = 150

• CHO/Pro = Multiply the dietary units by 1
  – 37.5 x 1 = 37.5

• Protein: determined by RDA
  – 5 year old = 1.1g/kg x 17.7 = 19.5g/day

• Carbohydrates: Obtained by subtracting protein amount above
  – 37.5 - 19.5 = 18 g carbohydrates
Diet Prescription

• 1500 calories/ day 4:1

  – 150 grams fat per day
  – 19.5 grams protein per day
  – 18 grams carbohydrates per day

  – This can then be divided into 3 meals and 2-3 snacks or 4 even meals
    • Replicate prior feeding regimen
Other Considerations for Diet Calculation

- Sugar alcohols
- Dietary fiber
- Carbohydrate from medications
Keto Admission

• JHH Admission Protocol
  – 4 families at a time
  – Admission 3 days (Monday-Wednesday)
  – Education Daily
  – Blood Glucose levels monitored
  – Urine ketone levels
    • Blood ketone levels

• Typical process:
  – Initiate KD slowly:
  – 24 hour fast (optional, but usually done)
  – Goal ratio at 1/2 calorie strength for 24 hours
    • Eggnog or ketogenic formula until full strength
  – Advance to Full Strength
    • With actual food
# Daily Education

<table>
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<tr>
<th>Day</th>
<th>Topics Covered</th>
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<tbody>
<tr>
<td>Monday</td>
<td>MD review&lt;br&gt;Diet basics&lt;br&gt;Side Effects&lt;br&gt;Diet initiation schedule&lt;br&gt;Meet with floor team/nurses</td>
</tr>
<tr>
<td>Tuesday</td>
<td>RD: The basics of the ketogenic diet&lt;br&gt;Meal Plan Guidelines&lt;br&gt;Social worker meeting</td>
</tr>
<tr>
<td>Wednesday</td>
<td>Ketogenic Computer program&lt;br&gt;What to do when your child gets sick&lt;br&gt;Parent lecture&lt;br&gt;Weighing and measuring foods&lt;br&gt;Learning how to read recipes</td>
</tr>
</tbody>
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Potential Side Effects

• Constipation
• Poor growth
• Osteopenia/ osteoporosis
• Kidney stones
• Hyperlipidemia
• Vitamin and mineral deficiency
Table 5. Recommendations for aspects of a follow-up KD clinic

<table>
<thead>
<tr>
<th>Nutritional assessment (registered dietitian)</th>
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<tbody>
<tr>
<td>Obtain height, weight, ideal weight for stature, growth velocity, BMI when appropriate</td>
</tr>
<tr>
<td>Review appropriateness of diet prescription (calories, protein, and fluid)</td>
</tr>
<tr>
<td>Review vitamin and mineral supplementation based on dietary reference intake guidelines</td>
</tr>
<tr>
<td>Assess compliance to therapy</td>
</tr>
<tr>
<td>Adjust therapy if necessary to improve compliance and optimize seizure control</td>
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<table>
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<tr>
<th>Medical evaluation (neurologist)</th>
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<tr>
<td>Efficacy of the diet (is the KD meeting parental expectations?)</td>
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<tr>
<td>Anticonvulsant reduction (if applicable)</td>
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<tr>
<td>Should the KD be continued?</td>
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<table>
<thead>
<tr>
<th>Laboratory assessment</th>
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<tbody>
<tr>
<td>Complete blood count with platelets</td>
</tr>
<tr>
<td>Electrolytes to include serum bicarbonate, total protein, calcium, magnesium, and phosphate</td>
</tr>
<tr>
<td>Serum liver and kidney profile (including albumin, AST, ALT, blood urea nitrogen, creatinine)</td>
</tr>
<tr>
<td>Fasting lipid profile</td>
</tr>
<tr>
<td>Serum acylcarnitine profile</td>
</tr>
<tr>
<td>Urinalysis</td>
</tr>
<tr>
<td>Urine calcium and creatinine</td>
</tr>
<tr>
<td>Anticonvulsant drug levels (if applicable)</td>
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<tr>
<td>Serum β-hydroxybutyrate (BOH) level</td>
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<tr>
<td>Zinc and selenium levels</td>
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<tr>
<td>Renal ultrasound</td>
</tr>
<tr>
<td>Bone mineral density (DEXA scan)</td>
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<tr>
<td>EEG</td>
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*aVisits should be at least every 3 months for the first year of the KD.*
Follow-up

• Generally every 3 months for the initial year
  – Infants seen in 1 month
  – Labs at each visit
    • CMP, Fasting lipid panel, urinalysis, Vit D, selenium
• Medications usually not changed the first month
• Frequent phone and email contact in between clinic visits
• At our center, all management through the keto team from now on...
Summary

• Ketogenic diet involves time and dedication on the dietitians part
  – But not difficult

• Requires a team
  – Physician, RN, Pharmacy support...
References

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