Creative Management of the Ketogenic Diet and Diet Therapies

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Lurie Children’s Epilepsy Center
The Ketogenic Diet Program

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The Dravet Syndrome Foundation Symposium
Lurie Children’s Ketogenic Diet Program

• Full time RD for Epilepsy Center
• Current caseload for KD: 160 patients
  Caseload in 2005: 15 patients
• The Keto Team
  Nurse Practitioners (Breanne Fisher, Catie Dezort)
  Social Worker (Josephine DeLira)
  Nutrition support (Wesley Lowman, Robyn Blackford)
• Diet therapy for epilepsy is a choice in treatment
• It is our passion to help families be successful on the diet by supporting them prior to the start of the diet, during the initiation of the diet and the day to day happenings in the future.
Types of Diet Therapies

• Ketogenic Diet (KD)
  – Classic (more long chain fats – butter, cream, oil)
  – MCT (more medium chain fats – MCT oil)

• Modified Atkins Diet (MAD)

• Low-Glycemic Index Treatment (LGIT)
The Ketogenic Diet

• Used for treatment of pediatric epilepsy

• High fat, low CHO, adequate protein

• Ketosis = burn fat for energy instead of carbohydrates, measured in urine/blood

• A proven therapy for the treatment of pediatric epilepsy
The KD ratio

Grams of Fat : Grams of Carbohydrate + Grams of Protein

Common ratio is 4:1

That means the KD can be about 90% fat and 10% Carb + Protein

Totaling about 2-10 grams Carb per day
Example of the KD ratio

Avocado

Grams per serving = 100
Grams of fat = 15
Grams of carb (-fiber) = 2
Grams of protein = 2

Ketogenic Ratio
Fat : Carb + Pro

Avocado Ratio
15 : 2 + 2
4 : 1
Example of KD ratio

Most foods are not a 4:1 ketogenic ratio; therefore, you have to add foods together to create the correct ratio.
Comparison

Regular/General Diet

Ketogenic Diet 4:1 ratio

Photos courtesy of Beth Zupec-Kania, RDN  Charlie Foundation
## Creating the Meals

<table>
<thead>
<tr>
<th>PROTEIN</th>
<th>CARB</th>
<th>CREAM</th>
<th>FAT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eggs</td>
<td>Lettuce</td>
<td>Cream</td>
<td>Butter</td>
</tr>
<tr>
<td>Bacon</td>
<td>Tomato</td>
<td>Whipped Cream</td>
<td>Coconut Oil</td>
</tr>
<tr>
<td>Sausage</td>
<td>Squash</td>
<td>Ice Cream</td>
<td>Margarine</td>
</tr>
<tr>
<td>Chicken</td>
<td>Broccoli</td>
<td>Cocoa</td>
<td>Mayo</td>
</tr>
<tr>
<td>Pork</td>
<td>Carrots</td>
<td>Sherbet</td>
<td>Oils</td>
</tr>
<tr>
<td>Beef</td>
<td>Green Beans</td>
<td>Popcicle</td>
<td></td>
</tr>
<tr>
<td>Fish</td>
<td>Applesauce</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cheese</td>
<td>Strawberries</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hot Dogs</td>
<td>Peach</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Calculating a Keto Meal

**Options:**
- Return to Main Menu
- Also Back to List
- Print or Email This Meal
- Delete Meal

**KETO CALCULATOR - Meal Edit**

**Meals for Ima Beginner**

**Important Information Regarding Meals:**
1. Be sure to check the verified box when you are satisfied with the meal. Only verified meals can be printed.
2. You must press the 'SAVE' button after changing any field or checking that the meal has been verified.
3. Tabbing to the next field will perform the calculation for fat, protein, carbohydrate and calories.

**Meal Created:** 9/23/2010 **Verified?** 

**Meal Name:** lunch

### Add New Foods

<table>
<thead>
<tr>
<th>Delete?</th>
<th>Food Item</th>
<th>Grams</th>
<th>Fat</th>
<th>Pro</th>
<th>Carb</th>
<th>Calories</th>
<th>Units</th>
<th>Ratio</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Cream, 36%</td>
<td>31</td>
<td>11.16</td>
<td>0.62</td>
<td>0.93</td>
<td>107</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Apple, raw - without skin</td>
<td>27</td>
<td>0.04</td>
<td>0.07</td>
<td>3.09</td>
<td>13</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Fish, Tuna Solid Wh. Albacore/water</td>
<td>12</td>
<td>0.38</td>
<td>3.04</td>
<td>0</td>
<td>16</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Mayonnaise, Hellmann’s/Best Foods</td>
<td>15</td>
<td>12</td>
<td>0.01</td>
<td>0.1</td>
<td>108</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Actual**
- Calories: 23.58
- Fat: 3.74
- Protein: 4.12
- Carbohydrates: 244
- Units: 7.86
- Ratio: 3.1

**Recommended**
- Calories: 23.71
- Fat: 3.6
- Protein: 4.3
- Carbohydrates: 245
- Units: 7.9
- Ratio: 3.1

Suggested accuracy: achieve within 4 calories of recommended calories AND on or slightly above the ratio.

**Meal Preparation Instructions**
## Example Keto Meals

<table>
<thead>
<tr>
<th>Breakfast</th>
<th>Lunch</th>
<th>Dinner</th>
</tr>
</thead>
<tbody>
<tr>
<td>60 gm Cream</td>
<td>60 gm Cream</td>
<td>60 gm Cream</td>
</tr>
<tr>
<td>31 gm Strawberries</td>
<td>29 gm Applesauce</td>
<td>42 gm Carrots (C)</td>
</tr>
<tr>
<td>28 gm Raw Egg</td>
<td>15 gm Lettuce</td>
<td>12 gm Chicken</td>
</tr>
<tr>
<td>15 gm Butter</td>
<td>14 gm Tuna</td>
<td>17 gm Butter</td>
</tr>
<tr>
<td></td>
<td>Make tuna salad</td>
<td></td>
</tr>
<tr>
<td></td>
<td>lettuce roll up</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Make scrambled eggs</td>
<td></td>
</tr>
</tbody>
</table>

2yr 8mo – male – IBW:13kg – 1200kcals/d – 3.5:1 KD ratio
Per Meal: 36.15gm Fat – 5.2gm Pro – 5.13gm CHO – 367kcal
Plus 100kcal snack
Example Meals

www.ketocalculator.com
Measuring Urine Ketones

• For the KD, it is typical to have moderate to large urine ketones.

• Most children are tested twice per day for ketone levels.
Tube and bottle feeds

• Initiation
  – Infants and tube feeds:
    • Mix Ketocal + formula (or BM)
    • Achieve correct ratio

• Transition from bottle to solids
  – Mix cream/whole milk with formula
  – Reduce formula while increasing solid foods

• Emergency situations
  – When the oral fed child urgently needs NG feeds inpatient

• Cooking and baking
  – Substitutes as flour in some recipes
Blenderized feeds and Purees

**Blenderized**

- For those that want to use “real food” vs formula
- Replace 1 tube feed with a blended mixture of foods per day to begin
- Will need a high powdered blender (VitaMix, Blendtec, etc)

**Purees**

- For those that want to start infants on solids or give tastes to the tube fed child
  - Oil in tube & food in mouth
- Infants can start normal progression and introduction of solid foods while on KD
- Keto RD can work with Speech Therapist for proper consistencies of foods for child
KD Initiation Process

MD identifies candidate for KD based on:
* diagnosis
* treatment plan
* parent/caregiver interest

Referral from MD to Keto Team for 1 hour outpatient appt
Lurie Children’s Ketogenic Diet Process

1. Meet our team & learn more about the diet.  
   - One hour appointment

2. Stay at the hospital & start the diet.  
   - Four day stay

3. Follow-up & answer any questions.  
   - One month after hospital stay

4. Ongoing support & counseling.  
   - Ongoing and as needed
KD Initiation Process

Initial Keto Team appointment:
- Go over handouts of the process to start KD
- Inquire about families’ worries/fears
- Find out about family structure and siblings
- Who takes care of the child during the day
- Ask about religious affiliations and special holidays or gatherings where there might be food
- What do they hope to gain by starting the KD?
Inpatient KD Initiation

- Day #1 KD 1:1 ratio at 1/3 kcals
- Day #2 KD 2:1 ratio at 2/3 kcals
- Day #3 KD GOAL 3:1 ratio at full kcals
- Day #4 same 3:1 ratio, go home

  - For infants and GT: no kcal restriction
Nutrition Education for KD Initiation Inpatient

Day 1: meet & greet and answer emergency questions and concerns; general guidelines

Day 2: sick day guidelines, menu preview, exchange list, cream explanation, scale

Day 3: Menus, KetoCalculator, free food list

Day 4: Supplements, fluid guide, wrap up
Polytherapy: Diet & AEDs & Supplements

• Medications must be in lowest carbohydrate form possible
  – Usually means tablets that can be crushed
  – All medications checked for carb, not just sugar
  – Possible decrease in AEDs at start of KD

• Supplements
  – Multivitamin
  – Calcium + vitamin D
  – Others: carnitine, magnesium, zinc, potassium, omega-3
Experience at Lurie Children’s: Side Effects

Why we might stop the KD (or maybe just lower the ratio)

- It's not working
- Behavior issues
- Family request
- Other treatment options that don't jive with KD
- Side effects
  - Hyperlipidemia
  - Metabolic Acidosis
  - Kidney Stones
  - Constipation
Other diets

- **Modified Atkins Diet**
  - Protocol by The Johns Hopkins Hospital
  - Initiate at 10 grams Carb (~1:1 ratio KD)
  - No restriction on calories, protein, or fluid
  - Children can often increase to 20 grams of carb after the first few months
  - Usually in ketosis
  - Diet started at home with close medical team supervision
Other diets

- Low Glycemic Index Treatment (LGIT)
  - Protocol by Massachusetts General Hospital in Boston
  - 40-60 grams of total carb
  - Carb sources <50 on the glycemic index
  - Usually not in ketosis, but have stable, lower blood sugars
  - Diet started at home with close medical team supervision
My Experience with Dravet Syndrome and the KD

- Present with thin, petite appearance
  - GOAL: Progress toward ideal body wt while on KD
- Can become ill and/or go into status epilepticus
  - GOAL: Create a long snack list as meal alternative
- Sensitive to changes in calories and ratio
  - GOAL: Make calorie changes in small increments
  - GOAL: Start at 3:1 ratio, ramp up slow, ¼ changes
- Periods of seizure freedom
  - GOAL: Flexible adjustment of the diet
Lurie Experience with Dravet Syndrome and the KD

Table 1. Twenty Patients With Dravet Syndrome Treated With the Ketogenic Diet.

<table>
<thead>
<tr>
<th>SCN1A</th>
<th>Failed seizure medications prior to diet</th>
<th>Age diet began (years)</th>
<th>Ketogenic diet ratio</th>
<th>Side effects</th>
<th>Impact on seizures</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Nonsense</td>
<td>7</td>
<td>2.8</td>
<td>2.75:1</td>
<td>Acidosis</td>
<td>&gt;50% GTC</td>
</tr>
<tr>
<td>2 Splice site</td>
<td>5</td>
<td>10.4</td>
<td>4:1</td>
<td>Constipation</td>
<td>&gt;50% GTC Resolved A, M</td>
</tr>
<tr>
<td>3 Nonsense</td>
<td>2</td>
<td>1.4</td>
<td>3:1</td>
<td>Acidosis</td>
<td>&gt;50% GTC, HC</td>
</tr>
<tr>
<td>4 Missense</td>
<td>5</td>
<td>2.0</td>
<td>4:1</td>
<td>Acidosis kidney stones</td>
<td>Resolved A, M</td>
</tr>
<tr>
<td>5 Missense</td>
<td>4</td>
<td>1.6</td>
<td>4:1</td>
<td>Acidosis compliance</td>
<td>&lt;50% GTC</td>
</tr>
<tr>
<td>6 Frameshift</td>
<td>7</td>
<td>2.3</td>
<td>4:1</td>
<td>None</td>
<td>&gt;90% A, M</td>
</tr>
<tr>
<td>7 Frameshift</td>
<td>6</td>
<td>3.7</td>
<td>2.75:1</td>
<td>None</td>
<td>&gt;90% GTC, M</td>
</tr>
<tr>
<td>8 Missense</td>
<td>5</td>
<td>1.3</td>
<td>3.75:1</td>
<td>Acidosis</td>
<td>&gt;50% GTC, M</td>
</tr>
<tr>
<td>9 Missense</td>
<td>6</td>
<td>3.9</td>
<td>4:1</td>
<td>Compliance</td>
<td>&lt;50% GTC, M</td>
</tr>
<tr>
<td>10 Nonsense</td>
<td>4</td>
<td>2.9</td>
<td>3:1</td>
<td>None</td>
<td>&gt;90% GTC, M</td>
</tr>
<tr>
<td>11 Frameshift</td>
<td>5</td>
<td>2.5</td>
<td>4:1</td>
<td>None</td>
<td>&gt;90% GTC, A, M</td>
</tr>
<tr>
<td>12 Splice site</td>
<td>5</td>
<td>6.1</td>
<td>4:1</td>
<td>None</td>
<td>&gt;50% GTC, M</td>
</tr>
<tr>
<td>13 Deletion</td>
<td>5</td>
<td>1.7</td>
<td>3:1</td>
<td>Compliance</td>
<td>&lt;50% GTC</td>
</tr>
<tr>
<td>14 Nonsense</td>
<td>5</td>
<td>1.5</td>
<td>4:1</td>
<td>Acidosis</td>
<td>&lt;50% GTC, M</td>
</tr>
<tr>
<td>15 Missense</td>
<td>9</td>
<td>6.3</td>
<td>4:1</td>
<td>Compliance</td>
<td>&lt;50% GTC, HC, M</td>
</tr>
<tr>
<td>16 Missense</td>
<td>6</td>
<td>2.7</td>
<td>3.25:1</td>
<td>None</td>
<td>50% GTC, A, M</td>
</tr>
<tr>
<td>17 Frameshift</td>
<td>5</td>
<td>3.9</td>
<td>4:1</td>
<td>None</td>
<td>&gt;50% GTC, A, M</td>
</tr>
<tr>
<td>18 Frameshift</td>
<td>5</td>
<td>3.5</td>
<td>3:1</td>
<td>None</td>
<td>&gt;50% A, M</td>
</tr>
<tr>
<td>19 Missense</td>
<td>5</td>
<td>4.4</td>
<td>3.5:1</td>
<td>None</td>
<td>&lt;50% GTC</td>
</tr>
<tr>
<td>20 Missense</td>
<td>7</td>
<td>2.9</td>
<td>3.25:1</td>
<td>Compliance</td>
<td>&lt;50% GTC, M</td>
</tr>
</tbody>
</table>

Abbreviations: A, atypical absence seizures; GTC, generalized tonic clonic seizures; HC, hemiconvulsions; M, myoclonic seizures.

The Ketogenic Diet in Dravet Syndrome
Linda Laux and Robyn Blackford
*J Child Neurol* published online 7 May 2013
DOI: 10.1177/0883073813487599
Lurie Experience with Dravet Syndrome and the KD

- 130 patients with DS reviewed
  - 56 patients (43%) had been on KD at some time
  - 48 families reported that KD had some benefit

- Of the 56 patients, 20 patients followed at Lurie on KD
  - All had a confirmed SCN1A mutation
  - Avg # medication trials before starting KD: 5.4 (range 2-9)
  - Avg age at diet initiation: 2.9 years (range 1.4-10.4 yrs)
Lurie Experience with Dravet Syndrome and the KD

- 13/20 (65%) had >50% reduction in sz
- 6/20 (30%) had >90% reduction in sz

- Reduced frequency of all sz types: generalized convulsions, hemi-convulsions, atypical absence, myoclonic sz

- 15/20 (75%) Parents reported:
  - increased alertness
  - improved cognition
  - improved behavior
Diet Therapy

Resources

• Lurie Children’s website – search for epilepsy, ketogenic diet
  – www.luriechildrens.org
• Epilepsy Foundation of Greater Chicago
• Myketocal.com
• Ketocook.com
• Modifiedmom.com
• Charlie Foundation website
• Matthew’s Friends website
Thank you!

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