The Ketogenic and Modified Atkins Diet for the Treatment of Epilepsy

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### What is the Ketogenic Diet?

- A high fat, low carbohydrate diet
- FDA approved treatment for Intractable Epilepsy
- Proven to be helpful for seizures that cannot be controlled with medication(s)



# History

- Throughout history it has been recognized that if a person with epilepsy stops eating their seizures will generally stop
- 1910 First scientific study on fasting is done in France
- 1921 Dr. Wilder at the Mayo Clinic first used the Ketogenic Diet and it was widely used until effective medications were introduced
- 1980's Dr. Freeman at John Hopkins Hospital revived the diet



#### How does it work?

- The body converts fat to energy instead of using carbohydrates
- Ketone bodies are produced as a by-product of fat metabolism
- "Keto" = ketone + "genic" = producing
- Ketones have anticonvulsant, sedative, and appetite suppression effects



# Description of the diet

- 90% of the total calories are provided from fat, therefore the meals are small due to the high fat content of the diet
- Remainder of calories are provided from protein and carbohydrate combined (protein intake is calculated using the DRI for age, to meet minimum recommended needs)
- Daily carbohydrate intake generally ranges between 2-5 grams per day.



#### **Diet Ratios**

#### Diet prescribed in ratios:

#### 4:1, 3:1, 2:1, 1:1



#### **Diet Ratios**

What do the numbers mean?

- Diet ratio is the dietary unit composition or building block of the diet
- It explains the composition of the diet
- For example a 4 :1 diet consists of 4 grams of fat for every 1 gram of carbohydrate and protein combined





### Foods Groups

Four food groups of the Ketogenic Diet

- Protein: meat, fish, poultry, egg, or cheese
- Carbohydrate: fruits and vegetables
- Fat: butter, oil, margarine, cream cheese, sour cream, or mayonnaise
- 36% or 40% heavy whipping cream



### Meal Plans

Solid food meal plans

- Meat/fish/poultry, fruit/vegetable, fat, cream
- Cheese, fruit/vegetable, fat, cream
- Egg, fruit/vegetable, fat, cream



# Meal Plans

- Liquid Diet
  - Ketogenic tube feeding
    - Ross Carbohydrate Free (RCF) formula
      - Microlipid or Liquigen
      - Solcarb powder
      - Prescribed amount of water
    - Ketocal powder or liquid
      - Powder: 4:1 or 3:1 ratio
      - Liquid: 4:1 ratio, contains fiber, flavored or unflavored
      - Prescribed amount of water
    - Ketovie 4:1, 3:1, and Ketovie Peptide.

# Other components of the diet

- Snack (1 of the following is allowed/day)
  - 2 lettuce leaves or ½ cup chopped lettuce (iceberg lettuce)
  - 1 walnut
  - 1 macadamia nut
  - 3 filberts
  - 3 black olives
  - 1 brazil nut
  - 1 butternut
  - 2 pecans
  - 1 tablespoon sour cream

#### Free foods

• Pepper or spices as desired

- 1 diet, caffeine free soda per day
- 1 piece of Trident sugarless gum per day
- 1 frozen popsicle per day made by freezing sugar free Kool-Aid or Crystal lite
- Weak decaffeinated coffee or tea
- Sugar free Kool-Aid made with liquid sweetener
- Extracts like pure vanilla, chocolate, coconut, maple (up to 15 drops each meal).
- Artificial liquid sweeteners without calories such as Sweet 10 or Sweet-N-Low

### Side Effects

Hypoglycemia

Osteoporosis

Constipation

Hyperacidosis

- Slows growth/weight
- Hyperlipidemia

- Kidney stones
- Pancreatitis



# Hypoglycemia

- Can be a problem for active persons
- The family is taught the s x s
  - Mild Irritable, pale, weak, drowsy, headache
  - Moderate Confusion, tremors, sweaty, increased pulse, feel cold and clammy
  - Severe Loss of consciousness
- Treatment 30cc juice or regular soda
  - Could use teaspoon of sugar, ice cream, pudding
- Important to eat several small meals per day
  - 4-6 meals



### Constipation

- It is seen in almost everyone on the diet
- Caused by decreased fiber in the diet
- Concurrent use of antiseizure medications
- Treatment
  - Smooth Move tea
  - SF MOM
  - Miralax

### Slows Growth/Weight

- Weight loss
- Growth slowed due to decreased amount of protein
  - If consume < 80% of calories or daily protein
- Monitoring by dietitian essential



# Hyperlipidemia

- Diet high in fats
- Familial predisposition
- Inability to metabolize fat efficiently
- Treatment
  - Adjust the diet
  - Carnitine



#### Osteoporosis

- Deficient in Calcium, Vitamin D, Magnesium, Phosphorous, Folic acid, and other trace vitamins and minerals
- Concurrent use of medications that interfere with Vitamin D absorption (Dilantin, Phenobarb, Tegretol)
- Many patients are non-weight bearing

Treatment – all patients given calcium and vitamin D supplements

### Hyperacidosis

- Ketones are acids and if they get too high they can cause hyperacidosis
- Ketone levels may be lower on MAD
- The body quickly compensates to correct its pH
- May occur when ill or dehydrated
- Treatment
  - Rehydrate
  - Adjust diet
  - May need to give Bicitra

# **Kidney Stones**

- Caused by lack of fluids
- Meds
  - Topiramate
  - Zonisamide
- Symptoms
  - Back pain
  - Difficulty with urination
  - Blood in the urine
- Treatment
  - Insure adequate fluid intake
  - Bicitra

#### Pancreatitis

 Cause is due to the pancreas inability to handle the increase of fats

Symptoms

- Severe abdominal pain
- Abdomen very tender to touch
- Vomiting
- Treatment
  - Discontinue the diet
  - Can never be restarted

# **Pros and Cons**

#### Pros

- More effective than medication in decreasing seizures
  - 75 % of patients will have 50% or more reduction
  - 27% of patients will have at 90% or more reduction
- Eliminate seizures in 10–15%
- Decrease medications
- Works well with infants and GT fed children

#### Cons

- Must weigh everything
- More difficult for adults
- Family lifestyle change
- Cultural lifestyle change
- Become bored with foods
- Tendency to "cheat"



# Lifestyle Changes

#### Monitor everything that is put in mouth

- Medications including IV fluids
- Over the counter meds cough syrups, pain meds
- Toothpaste
- Sunscreens and lotions
- Record dietary intake



### Monitoring ketone levels

#### Educate family on Ketostix

- Urine collection
- Reading Ketostix



#### Daily for 7-10 days then as needed





#### Duration of diet therapy

- 4:1 diet for 2 years until "seizure-free" w/o meds for 1 full year
- 3:1 diet for 6 months, if "seizure free"
- > 2:1 diet for 6 months, if "seizure free"
- No concentrated sweets diet for 6 months post d/c of diet



### Prior to clinic appointment

- RN discusses diet with team
- What are expectations
- Lifestyle change for most families
- Suggest decreasing carbohydrates
- > 2–3 month commitment

#### Preadmission

- Insurance Prior Authorization
  - Hospitalization–usually 3–5 days
  - Specialized Formula if needed
  - Medications cannot contain carbohydrates
- Equipment
  - Scale
- Baseline lab work
- Education
  - *Everything* must be checked for carbohydrate content



### Hospitalization

- Patients to arrive at 0900
  - Have been fasting since the night prior to admission
  - If they are not on the floor by 1000 please call admitting
- Monitor blood sugar levels every 6 hours
  - Do not reach nadir until BS at ~ 40
  - Will usually rise again into 60's 70's
- Monitor I & O and urine ketones
- Monitor for lethargy
- Monitor toleration of increased fats



### How is the diet started?

- Patient is admitted for 3–5 days
- Diet is started by gradually increasing fat intake to the goal amount and by decreasing the carbohydrate intake
- Outpatient
  - High fat, low carbohydrate diet transitioned slowly and gradually over 2-3 months depending on the patients current diet (oral versus enteral fed)



#### How is the diet started inpatient?

- Day 1: carbohydrate intake is decreased significantly and fat intake is provided as 30% of the total calories
- Day 2: carbohydrate intake remains at the amount provided on Day 1 and fat intake is provided as 60% of the total calories
- Day 3: carbohydrate intake remains at the amount provided on Day 1 and fat intake is provided as 90% of the total calories

# Solid Food Diet

- Day 1 and Day 2: liquid diet, usually in the form of Ross Carbohydrate Free formula with Microlipid and Polycose powder; or Ketocal 4:1 or Ketocal 3:1 formula
  - Patient is offered 3 meals per 24 hours
  - Ideally meals should be offered on a schedule as the diet is utilized as a medication, and because a steady intake of glucose is required to maintain blood glucose levels

Day 3: Start solid food meals

#### Formula Diet

- Day 1 Formula: provide 30% of calories from fat and decrease carbohydrate intake
- Day 2 Formula: provide 60% of calories from fat and decrease carbohydrate intake as prescribed for Day 1
- Day 3 (Goal Formula): provide 90% of calories from fat and decrease carbohydrate intake as prescribed for Day 1



#### Education During the Admission

- Day 1: discuss with parents and patients progression of the diet over the course of the hospital stay
- Day 3: Meet with parents to provide instructions on preparation of formula and/or solid food meals.
  - How to recognize s/s of hypoglycemia
  - How to treat hypoglycemia
  - Review OTC medications that can be utilized with the diet

# Monitoring During Initiation of the Diet

- Initial labs
- Monitor overall tolerance of diet (vomiting and/or diarrhea)
- Monitor blood glucose levels and urine ketones



### Lab Monitoring

- Baseline lab work prior to starting the diet
- Every 3–6 months the first year
- Every year the second year
- Ketogenic Diet Panel
  CBC, Chem7, Lipid Panel, PO4, MG, Prealbumin, HFP, Vitamin D
   Medication Levels


### **Ongoing Monitoring**

- Follow up 2 weeks after starting the diet
- Urine ketone testing at home as necessary
- Every 3 months thereafter in clinic
  - Weight/height check
  - Assess for side effects
  - Ongoing education
- Keto Panel



# Ketogenic diet may also help

- Diabetes
- Amyotrophic lateral sclerosis (ALS)
- Alzheimer's
- Parkinson's disease
- Some mitochondriopathies (carnitine deficiency)
- Glut 1 deficiency
- TBI (Strokes)
- Cancer (gliomas)
- Autism



#### **Modified Atkins Diet**

- Limited to ~ 20 grams carbs/day
  - ~ 65% of calories are provided from fat
- Modified refers to the lower carbohydrate limit compared to Atkins recommendations, with an emphasis on high fat foods
- Diet was created at Johns Hopkins to offer a less restrictive diet primarily for teenagers and adults
  - Easier diet for patients who eat by mouth



## **Modified Atkins Diet**

- Usually 3 meals, 3 snacks
- Foods are measured, not weighed
  - Offers flexibility
  - Easier to be in social outings, restaurants
- Deficient in calcium, phosphorus, magnesium, zinc, folic acid. Vitamin and mineral supplements are recommended
- Monitored by team (MD, RN, RD)

#### When is the diet used?

- As an introduction to a carbohydrate restricted diet or as a trial to see if patient will respond to dietary therapy
- To transition off the Classical Ketogenic Diet
- Families with limited time and resources
- Older children, teenagers, adults



## Efficacy

#### Efficacy

- ~ 66 percent of children following the Modified Atkins Diet at Johns Hopkins had a 50% reduction in seizure activity
- ~ 45% of patients who have trialed the diet will show improvement in seizure control



# Lifestyle Changes

- Many cultures socialize around food/eating
  - Dining out
  - Special occasions birthdays, holidays
- Family must be willing to monitor closely
- Easier if high carb foods not in house
- Cheating happens



#### Prior to clinic appointment

- RN discusses diet with team
- What are expectations
- Lifestyle change for most families
- Suggest decreasing carbohydrates
- > 2–3 month commitment

#### **RN** Consultation

- RN provides general diet education on how to decrease carbohydrate and increase fat intake until the patient is seen in clinic
- Suggest ways to decrease carbohydrates; omit concentrated sweets, breads, pastas, dairy, limit fruits and starchy vegetables
- > 2–3 month commitment



# **Modified Atkins Diet Education**

- Detailed education about food groups
  - Fat is emphasized
    - Heavy cream, butter, oils, cream cheese, sour cream
  - Protein is not limited
    - Meat, chicken, fish, turkey, cheese, eggs, nut butters
  - Carbohydrates are limited to lower carbohydrate fruits and vegetables
    - Berries, olives, broccoli, peppers, cauliflower, celery, cucumber, mushrooms, romaine lettuce, spinach, zucchini, avocados, spaghetti squash



## **Reading Food Labels**

- Educate primary caregiver(s) how to read food labels
  - School
  - Other family members
  - Friends
- Count "Total Carbohydrates"
- Pay attention to portion sizes
- Divide 20 grams of carbs throughout the day



# **Ongoing Monitoring**

- ▶ Follow up: 4–6 weeks after starting the diet
- Urine ketone testing at home as necessary
- Every 3 months thereafter in clinic
  - Weight/height check
  - Assess for side effects
  - Ongoing education
    - Lots of phone calls and support for patients who are new to the diet

Keto Panel

#### Sample menu for the Modified Atkins Diet

- Breakfast
  - Egg Scramble
    - 2 large eggs
    - 2 Tbsp heavy cream
    - 1 Tbsp butter
    - 1/4 cup feta cheese
    - 1/2 cup spinach
    - 1/2 cup mushrooms, chopped

- Lunch
  - Cobb Salad
    - 1 1/2 cups mixed greens
    - 1/2 cup avocado, sliced
    - 1 hard-boiled egg, sliced
    - 1 Tbsp finely chopped bacon
    - 1/4 cup blue cheese or cheddar cheese, shredded
    - 2 Tbsp olive oil
    - 1 Tbsp red wine vinegar

# Sample menu for the Modified Atkins Diet

- Dinner
  - Chicken and Zucchini "Pasta"
    - 1 medium baked chicken breast
    - 1 cup sliced or spiraled zucchini
    - 1 Tbsp. olive oil
    - 2 Tbsp. pesto



#### Sample Snack for the Modified Atkins Diet

- Snacks
  - Celery & Cream Cheese
  - 1 stalk of celery, sliced
  - 2 Tbsp full-fat cream cheese
  - Sugar-Free Gelatin, 1/2 cup



#### **Classical Keto & MAD**



#### Summary: Differences and Similarities

#### Differences

- Modified Atkins Diet
  - Protein not restricted
  - No gram scale required
  - More flexibility
  - Outpatient initiation
- Similarities
  - Classic Ketogenic Diet and Modified Atkins Diet
    - Carbohydrates are restricted
    - Fat is encouraged
    - Supplements are required
    - Both are effective with helping to manage seizures

#### Ketogenic Diet Program

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