

# **Chewing the Fat on the Nutritional Management of FODs**

**2006 FOD/OAA National Metabolic  
Family Conference**

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**Ross Metabolics**

# Fatty Acid Oxidation Disorders (FODs)

- **SCAD: Short-Chain Acyl CoA Dehydrogenase**
- **SCHAD: Short-Chain L-3 Hydroxyacyl Dehydrogenase**
- **MCAD: Medium-Chain Acyl CoA Dehydrogenase**
- **LCHAD: Long-Chain 3 Hydroxyacyl CoA Dehydrogenase**
- **VLCAD: Very Long-Chain Acyl CoA Dehydrogenase**

# FODs

- **Carnitine Disorders**
  - **Primary Carnitine Deficiency**
  - **Carnitine Update Defect (CUD)**
  - **Carnitine Acylcarnitine Translocase Deficiency (CAT)**
  - **Carnitine Palmitoyl Transferase (CPT)**
    - **CPT I**
    - **CPT II**

# FODs

- **Others**

- **Trifunctional Protein (TFP) Deficiency**
- **2,4 Dienoyl-CoA Reductase Deficiency**
- **Electron Transfer Flavoprotein (ETF) Dehydrogenase**
  - **Glutaric Aciduria Type II (GAll deficiency)**
  - **Multiple Acyl CoA Dehydrogenase Deficiency (MADD)**
- **3 Hydroxy-3- Methylglutaryl CoA Lyase (HMG) Deficiency**
- **Medium-Chain 3 Ketoacyl CoA Thiolase (MCKAT) Deficiency**

# Nutrition and FODs

- **Overview of nutrition consequences of FOD**
- **Nutrition management**
- **Controversies**
- **Your questions and concerns!**





# What Happens When We Eat?

## Carbohydrate



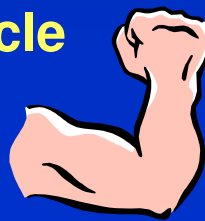
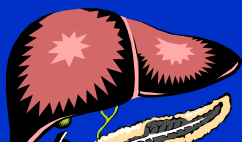
Glucose



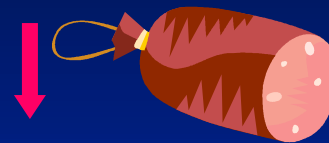
Glycogen



Liver Muscle



## Protein



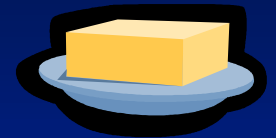
Amino Acids



Tissues



## Fat



Fatty Acids

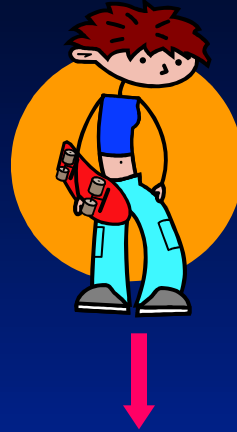


Heart

Adipose Tissue



# What Happens When We Fast?



**Glycogen**

**Protein**

**Fat**



**Amino Acids**

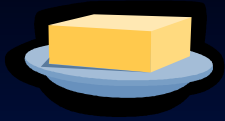
**Fatty Acids**



**Glucose**

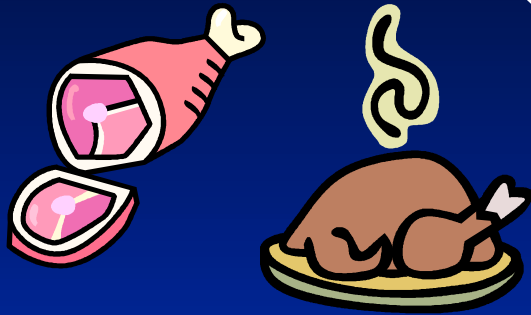
**Ketones**

# What Happens in FOD?



Protein + Fat in Food  
(GAI)

Protein Body + Fat in Body  
Stores



Fatty Acids + Amino Acids

Ketones

Lack of Energy

Growth  
development



Cardiomyopathy  
Arrhythmia

Muscle pain  
↓ tone



Sleepy  
↓ consciousness





# Symptoms of FODs

- **Hypoglycemia (low blood sugar)**
- **Non-ketotic (low ketone production)**
- **Acidosis**
- **Increased blood ammonia**
- **Toxic end products of fats**
- **Liver problems (↑ enzymes)**
- **Brain problems (altered consciousness, nervous system affects)**
- **Muscle problems (↓ tone, rhabdomyolysis)**
- **Eye problems (retinopathy)**

# What Happens in the Cell?

Long-chain fatty acids

+ Acyl CoA

Toxic Waste



CPT I

CPT II

VLCAD, LCHAD, MCAD, SCAD

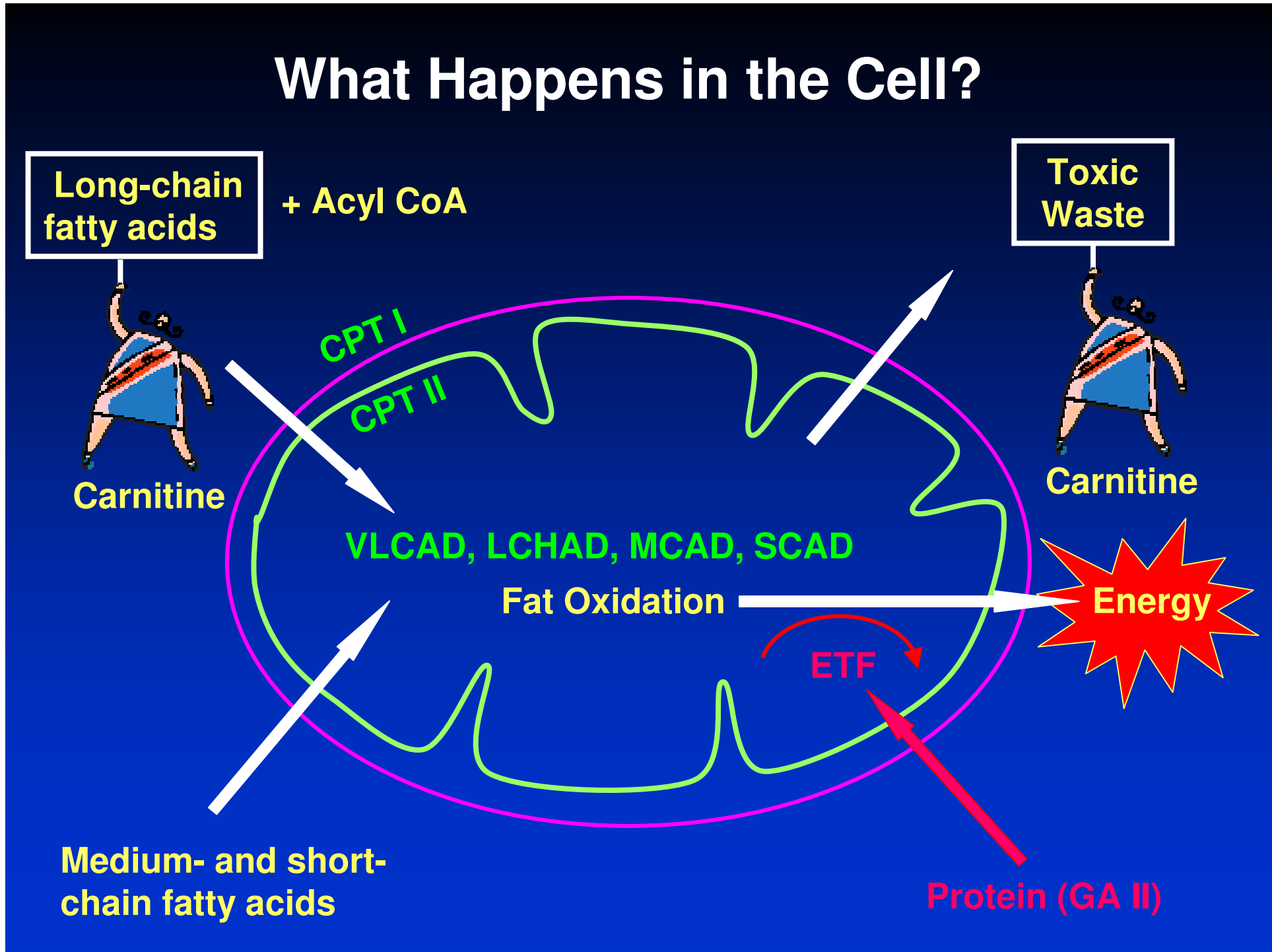
Fat Oxidation

Energy

ETF

Medium- and short-chain fatty acids

Protein (GA II)

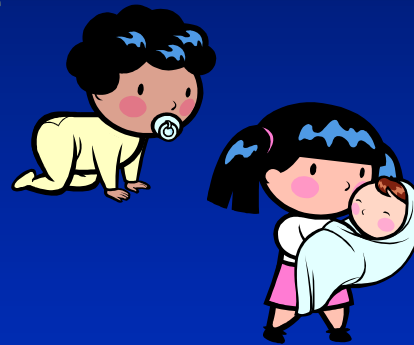


# Nutritional Treatment

- **AVOID FASTING !**
- **Fat restriction**
- **High carbohydrate for energy**
- **Protein restriction (GA II)**
- **Supplements**
  - **MCT oil**
  - **DHA (docosahexaenoic acid)**
  - **Carnitine**
  - **Riboflavin**
  - **Creatine monohydrate**

# Avoid Fasting !

- **Caloric intake MUST meet demand**
  - **Stress, illness, exercise**
- **Individual tolerance**
  - **Infants ~ 4 hrs**
  - **Children ~ 8 hrs**
- **Feeding**
  - **Continuous nighttime feeding**
  - **Cornstarch**



\* Remember: Fatty acids increase before blood glucose drops



## Fat Restriction - Why?

- **Toxic fatty acids may accumulate**
- **Excess fat may deplete carnitine**
- **Moderation of fat is key to a healthy diet**

# Fat Restriction - How Much?

- **MCAD, SCAD, CPTs, GA II**
  - **Total fat 25 - 30% of calories**
    - Necessary to restrict in infancy?
    - Important source of energy
  - **Restrict MCTs**
- **VLCAD, LCHAD**
  - **10 - 15% of calories from long-chain fats**
  - **Supplement with MCT (total 30 - 40%)**
  - **Provide essential fatty acids**

# Essential Fatty Acids

- **Body cannot produce**
- **Important for growth, skin, brain, infection, immunity**
- **Linoleic - 18:2, omega 6**
  - **4 to 15 g/day**
  - **3 to 5% of total calories**
- **Linolenic - 18:3, omega 3**
  - **0.5 - 1 g/day**
  - **1% of total calories**

# Sources of Essential Fatty Acids

Oil	1 Tbsp Linoleic (g)	1 Tbsp Linolenic (g)
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**Corn**

**8**

**0.1**



**Soy**

**7**

**0.9**



**Walnut**

**7**

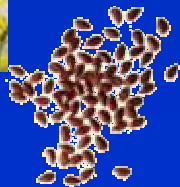
**1.4**



**Canola**

**3**

**1.5**



**Flaxseed**

**2**

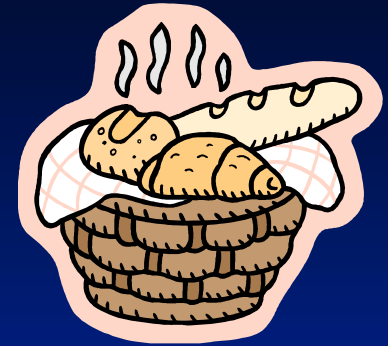
**7.0**



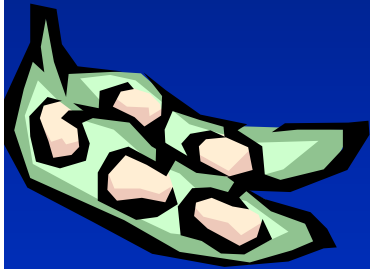
# Carbohydrate for Energy



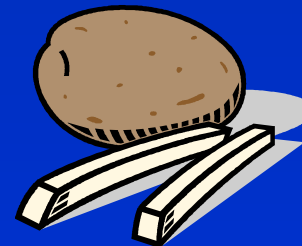
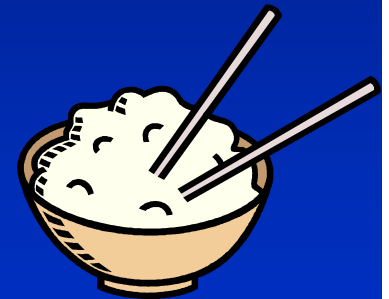
- Preferred energy source



- Complex carbohydrates - breads, cereals, vegetables



- Slowly digested for gradual glucose release



# Carbohydrate for Energy

- **Goal - Prevent Body Fat Breakdown**
- **Acute**
  - **IV glucose**
  - **Carnitine to help get rid of toxic by-products**
- **Chronic**
  - **Cornstarch**
    - **Nighttime feedings**
    - **Before exercise**
    - **During illness**



# Protein Restriction (GA II)



**Protein + Fat  
from Foods**

**Protein + Fat  
from Body Stores**

**Growth**

**Energy**



- **Cannot use fats and protein for energy**
- **Need enough protein for growth**

# MCT Oil

- Used for long-chain disorders

- Infant Formulas

- ProViMin + MCT + EFA



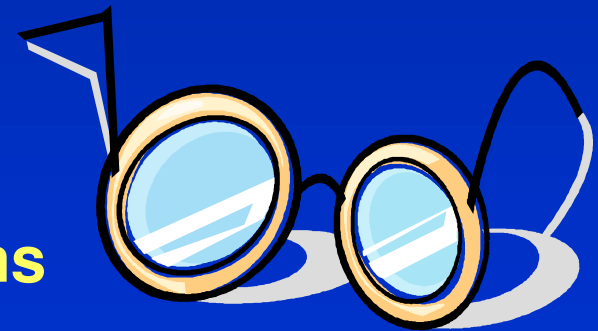
- Portagen + EFA



- Can use in cooking, salad dressings

# Docosahexaenoic Acid (DHA)

- **Very-long chain fatty acid (22:6)**
- **Found naturally in fish, shellfish, breastmilk**
- **Can be made from linolenic acid (18:3)**
- **DHA needed in infant development for brain and eye**
- **65 to 130 mg/day may be helpful for LCHADs**  
*(Gillingham et al 2003)*
- **May improve visual acuity**
- **Will not prevent retinal problems**



# Medium-Chain Triglyceride (MCT) Oil

- **Readily available energy source**
- **Reduces long-chain fatty acid oxidation**
- **Prevents build-up of toxic metabolites**
- **Improved metabolic control in LCHAD patients (*den Boer 2002*)**

# Carnitine

- **Transport of fatty acids into the mitochondria**
- **Conjugates toxic acyl fats for excretion**



# Carnitine - Pros



- **Corrects secondary deficiencies**
  - Patients with FOD **MAY** have low levels
- **Conjugates toxic fats for excretion**
- **MCAD - use during illness**
- **MCAD - improved exercise tolerance**





## Carnitine - Cons

- **Excess long-chain acylcarnitines *MAY* accumulate in tissues**
- **50 LCHAD patients - no improvement in frequency of metabolic decompensation (*den Boer 2002*)**
- **MCAD - no change in fasting tolerance**

# Riboflavin

- **Riboflavin part of FAD (flavin adenine nucleotide)**
- **Cofactor of SCAD enzyme and in the mitochondria**
- **GA II and SCAD - some patients riboflavin-responsive**



# Creatine Monophosphate

- Creatine is an energy source for muscle



- Used with carbohydrate to delay fat oxidation
- Role in exercise
- Improvement in LCHAD (*Shortland 2001*)

# Questions?



# What Happens If You Consume Too Much Fat?

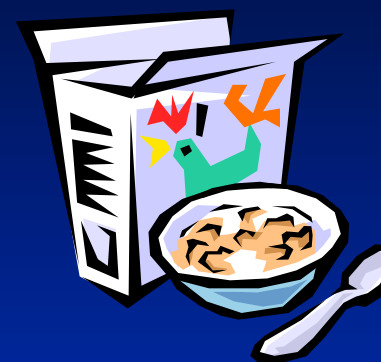
- **Excess fat storage?**
- **1 gram = 9 calories**
- **High-fat, low-carbohydrate**
  - **will need to use fat for energy**
- **Depletes carnitine**



# How Do You Keep at 25-30% Fat Without Counting Grams?

Nutrition Facts	
Serving Size 2 crackers (14 g)	
Servings Per Container About 21	
Amount Per Serving	
<b>Calories</b> 60	Calories from Fat 15
% Daily Value*	
<b>Total Fat</b> 1.5g	<b>2%</b>
Saturated Fat 0g	<b>0%</b>
Trans Fat 0g	
<b>Cholesterol</b> 0mg	<b>0%</b>
<b>Sodium</b> 70mg	<b>3%</b>
<b>Total Carbohydrate</b> 10g	<b>3%</b>
Dietary Fiber Less than 1g	<b>3%</b>
Sugars 0g	
<b>Protein</b> 2g	
Vitamin A 0%	• Vitamin C 0%
Calcium 0%	• Iron 2%
* Percent Daily Values are based on a 2,000 calorie diet. Your daily values may be higher or lower depending on your calorie needs:	
	Calories: 2,000 2,500
Total Fat	Less than 65g 80g
Sat Fat	Less than 20g 25g
Cholesterol	Less than 300mg 300mg
Sodium	Less than 2400mg 2400mg
Total Carbohydrate	300g 375g
Dietary Fiber	25g 30g

- **No easy method !**
- **Limit high-fat foods**
- **Emphasis on carbohydrates**
- **Familiar with food composition**



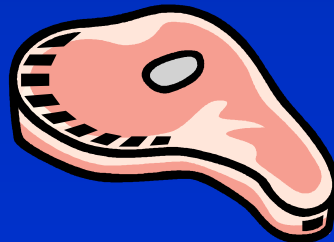
# Can a Child Have Fat Allowance All in One Meal?

- **Fat provides satiety**
- **Fat slower to digest**
- **Best to have both fat and carbohydrate in same meal**

***Snacking is important!***

# Vegetable vs Animal Protein

- Exclude **high-fat** protein choices
  - High-fat meat, dairy products
- Combination of both
- Can meet protein requirements on a vegetarian diet
- Animal protein excellent source of iron and zinc





# Compliance to the Diet

- **Parents may be less strict with discipline**
- **Parenting stress and anxiety**
- **Strong social support**



# You Can't Make Me Eat That!

- **Toddlers**

- Must be a consequence
- Attention seeking



- **Children**

- Increase understanding
- Encourage choices



- **Teens**

- Independence from the family
- Peer pressure





**Matthew**  
**VLCAD**

