Cystic Fibrosis and Nutrition

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Mountain State Cystic Fibrosis Center

Learning Objectives

• Classify age specific nutrition related guidelines for Cystic Fibrosis patients.
• Understand supplementation of fat soluble vitamins: A, D, E and K.
• Identify signs and symptoms of Pancreatic Insufficiency in patients with Cystic Fibrosis.
• Calculate pancreatic enzymes and make recommendations for Pancreatic Insufficient patients with Cystic Fibrosis.
• Recognize comorbidities related to Cystic fibrosis.

What is Cystic Fibrosis

• Cystic fibrosis (CF)
  • Chronic disease that affects the lungs and digestive systems of about 30,000 children and adults in the United States (70,000 worldwide)
  • A defective gene inherited from both parents which causes the body to produce unusually thick, sticky mucus.
  • Clogs the lungs and leads to life-threatening lung infections.
  • Obstructs the pancreas and stops natural enzymes from helping the body break down and absorb food.

• Sweat Test
  • Gold Standard
  • Newborn Screening
  • Improve growth
  • Help keep lungs healthy
  • Reduce hospital stays & Add years to life
  • Genetic Carrier Testing

Statistics

• About 1,000 new cases of cystic fibrosis are diagnosed each year.
  • More than 70% of patients are diagnosed by the age of two.
  • At least 45% of the CF patient population is age 18 or older.
  • The predicted median age of survival for a person with CF is in the late 30s.
  • In the 1950s, few children with cystic fibrosis lived to attend elementary school.

Symptoms of Cystic Fibrosis

• People with CF can have a variety of symptoms, including:
  • very salty-tasting skin
  • persistent coughing
  • frequent lung infections
  • wheezing or shortness of breath
  • poor growth/weight gain in spite of a good appetite & frequent greasy, bulky stools or difficulty in bowel movements
The Dietitian’s Key Roles in the Care of Cystic Fibrosis

• Monitor Absorption of Nutrients
• Identify Nutritional Status
• Provide Diet Education
• Assess Nutritional Needs
• Provide Enteral and Parenteral Nutrition Recommendations
• Assist in Recommendations of Pancreatic Enzymes

Energy and Macronutrient Guidelines

• Energy and Protein
  • Calorie Requirements: 1.2–2 times the DRI for age.
  • Protein Requirements: 1.5–2 times the DRI for age.
• Energy needs will be influenced by:
  • Severity of lung disease
  • Degree of malabsorption

Vitamins and Minerals

• Patients with CF do NOT absorb nutrients properly.
  • Increased need for fat soluble vitamins:
    • Vitamins A, D, E & K
    • CF Vitamins:
      • AquaDEX – Liquid, Chewable & Gel Capsule
      • SourceCF – Liquid, Chewable & Gel Capsule
    • Vitamins – Liquid & Chewable
  • Increased need for minerals
    • Calcium, Iron, Sodium Chloride & Zinc

Monitoring Serum Vitamin Levels

• At Diagnosis
  • Infants
    • Check vitamin levels 2–4 months after starting supplemental vitamins
  • Children/Adolescents & Adults
    • Check levels at diagnosis
• After Diagnosis
  • Vitamin A, D 25-OH, and E levels should be checked annually.
  • Vitamin K can be assessed using PIVKA II Or indirectly by using Prothrombin Time.
  • Possible K deficiency: easy bruising, difficulty with blood clotting.

Vitamin Recommendations

<table>
<thead>
<tr>
<th>Age</th>
<th>Liquid</th>
<th>Chewable</th>
<th>Soft-Gel</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-12 months</td>
<td>1 ml (0.5 ml BID)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1-3 years old</td>
<td>2 ml (1 ml BID)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4-8 years old</td>
<td>1 tablet</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9-18 years old</td>
<td>2 Capsules (1 Capsule BID)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&gt;18 years old</td>
<td>2 Capsules (1 Capsule BID)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

• Additional supplementation:
  • Vitamin A: 8,000–10,000 IU once daily
  • Vitamin D: 10,000–20,000 IU weekly
  • Vitamins with meals and enzymes

Signs of Malabsorption and Malabsorption

• Poor weight gain despite a good appetite.
• Frequent, loose and/or large bowel movements.
• Foul-smelling bowel movements.
• Mucus or oil in the bowel movement.
• Excessive gas and/or stomach pain.
• Distention or bloating.
**Pancreatic Sufficiency (PS) & Insufficiency (PI)**

- **Identifying PS or PI:**
  - Test 72 hour fecal
  - >200 Normal
  - 100 – 200 Moderate to Mild Exocrine PI
  - <100 Severe Exocrine PI

- Pancreatic Insufficient Patients
  - Prescribed Pancreatic Enzymes
- Pancreatic Sufficient Patients
  - Do not take enzymes

**What Are Enzymes & How Do They Work?**

- Pancreatic Enzyme Replacements
  - Lipase, Amylase & Protease
- Capsule Form
  - Inside each capsule are many small “beads” that contain digestive enzymes.
  - Each bead is covered with a special “coating.”
  - This coating allows the beads to dissolve in the small intestine.

- The main functions of enzymes are:
  - To digest carbohydrate, protein and fat
  - To help with weight gain &
  - To promote nutrient absorption.

**How Are Enzymes Given?**

- Enzymes should be taken:
  - Before meals and snacks.
  - Before, during and after nightly tube feeding.
- Older Children and Adults
  - Capsules should be taken with liquid and swallowed whole.
- Infants and Small Children
  - Capsules may be opened and beads can be mixed with a soft acidic food.
  - Applesauce

**FDA Approved Pancreatic Enzymes**

<table>
<thead>
<tr>
<th>Enzyme</th>
<th>Strengths (USP Units of Lipase)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Creon 24,12,6 &amp; 3</td>
<td>24000, 12000, 6000 &amp; 3000</td>
</tr>
<tr>
<td>Zenpep 25,20,15,10,5 &amp; 3</td>
<td>25000, 20000, 15000, 10000, 5000 &amp; 3000</td>
</tr>
<tr>
<td>Ultresa 23,20, &amp; 13</td>
<td>23000, 20700 &amp; 13800</td>
</tr>
<tr>
<td>Viokase 16 &amp; 8</td>
<td>16000 &amp; 8000</td>
</tr>
<tr>
<td>Pancrease 21,16,10 &amp; 4</td>
<td>21000, 16800, 10500 &amp; 4200</td>
</tr>
<tr>
<td>Pertzye 16 &amp; 8</td>
<td>16000 &amp; 8000</td>
</tr>
</tbody>
</table>

**Calculating Pancreatic Enzymes**

- **Dosing enzymes:**
  - Max - 2500 U Lipase/kg/meal
  - Max - 10,000 U Lipase/kg/day
- Infants and young children:
  - Start with 1,000 U Lipase/kg/meal until max dose is reached.
- Adults:
  - Start with 500 - 1,000 U Lipase/kg/meal until max dose is reached.

- **To calculate the enzyme dose per kg/meal:**
  - Multiply Units of Lipase by meal dose
  - Divide the total Units of lipase per meal by weight

- **Example:**
  - Enzyme prescription - Creon 12 (12,000 U of lipase) with 3 capsules per meal
  - Weight 15 kg
  - 12,000 x 3 = 36,000 U Lipase/meal
  - 36,000 divided by 15 kg = 2400 U Lipase/kg/meal
*Calculating Pancreatic Enzymes*

To calculate the enzyme dose per kg/day:
- Multiply Units of lipase by day dose
- Divide the total Units of lipase per day by weight

Example:
- Enzyme prescription - Creon 12 (12,000 U of Lipase) with 4 capsules per meal and 2 capsules per snack (3 meals and 2 snacks = 16 capsules per day)
- Weight 20 kg
- 12,000 X 16 = 192,000 U Lipase/meal
- 192,000 divided by 20 kg = 9,600 U Lipase/kg/meal

*Nutrition Through the Life Span*

- High-calorie diet, including supplements when needed.
- Behavioral intervention to encourage good eating habits in children.
- Keeping track of nutritional indicators, such as body mass index.
- Appropriate doses of pancreatic enzymes.

*Nutritional Management of Infants*

- Ages 0-12 months
  - Breast milk, iron-fortified formula
  - Enzymes prior to all feedings
  - Vitamin supplement
  - Salt supplementation
  - 1/8 tsp 0-6 months
  - 1/4 tsp 6-24 months
  - Add solids at 4 to 6 months
  - Referrals to community programs
  - WIC
  - Children with Special Health Care Needs

*Nutritional Management of Toddlers & Preschoolers*

- Ages 1-4
  - Provide a normal, healthy diet with a variety of high-calorie foods and calcium rich foods.
  - Encourage regular, pleasant meals and snacks.
  - Avoid “grazing” or constant snacking.
  - Teach appropriate self-feeding skills.
  - Continue vitamin, enzyme, and salt supplementation.

*Nutritional Management of Adolescents*

- Ages 12-17
  - Limit sweetened beverages.
  - Make recommendations for easy, quick, high-calorie foods.
  - Increase energy intake during growth spurts.
  - Promote independence with vitamin, enzyme, and salt administration.

*Nutritional Management of Adults*

- Ages 18 and Older
  - Well-balanced diet to support optimal nutritional status.
  - Sufficient calories to maintain healthy body weight.
  - Observe for Anorexic Behaviors.
  - Assess for Age-Related Complications of CF.
  - Continue Vitamins/Enzymes & Add Supplements as Needed.
**Identifying Nutritional Status**

<table>
<thead>
<tr>
<th>Red</th>
<th>Yellow</th>
<th>Green</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nutritional Failure</td>
<td>Nutritional Risk</td>
<td>Nutritional Health</td>
</tr>
<tr>
<td>Infants (0-23 months)</td>
<td>50th ht-wt</td>
<td>≥ 50th ht-wt</td>
</tr>
<tr>
<td>Children (2-20 yrs. of age)</td>
<td>10th-49th BMI for Age</td>
<td>≥ 50th BMI for Age</td>
</tr>
<tr>
<td>Adults (20 yrs. of age and older)</td>
<td>≤ 18 kg/m² for BMI</td>
<td>≤ 18 kg/m² for BMI Males</td>
</tr>
<tr>
<td></td>
<td>18-23 kg/m² for BMI Females</td>
<td>22 kg/m² for BMI Females</td>
</tr>
</tbody>
</table>

**Comorbidities Related to Cystic Fibrosis**

- Cystic Fibrosis Related Diabetes (CFRD)
- Cystic Fibrosis Related Liver Disease
- Cystic Fibrosis Related Renal Disease

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**Cystic Fibrosis Related Diabetes (CFRD)**

- Cystic Fibrosis-Related Diabetes (CFRD) is a unique type of diabetes
  - It is NOT the same as Type 1 or Type 2 Diabetes
  - 35% of adults 20 to 29 years of age
  - 43% of adults over 30 years of age
- Symptoms of CFRD
  - Increased thirst and increased urination.
  - Excessive fatigue, weight loss and unexplained decline in lung function.
- Screening and Diagnosis
  - Screening begins at 10 years of age
  - Hemoglobin A1C NOT a good indicator for diagnosis
  - 2 hour OGTT < 200 mg/dl
  - FBG > 126 mg/dl on two or more occasions
- Treatment of CFRD
  - Insulin
  - Keeping blood glucose levels at a normal or near-normal level

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**Cystic Fibrosis Related Liver Disease**

- More than 10 percent of people with CF have liver disease, a number that may increase as people with CF live longer lives.
- Maintenance of a "Normal" Nutritional State
- Preventing Deficiencies
- Protein and Fat Recommendations
- Depend on Severity of Disease
- Increased Energy Intake
- Fat Soluble Vitamins
- Monitor Every 6 to 12 Months
- Counseling Related to Risks of Alcohol Use

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**Cystic Fibrosis Related Renal Disease**

- Maintenance of a "Normal" Nutritional State
- Preventing Deficiencies
- Increased Energy Needs
- Protein and Salt Recommendations
- Depend on severity of the disease
- Monitoring of Renal Function Labs

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**Table 2. MNT For Type 1/Type 2 Diabetes Versus For CFRD**

<table>
<thead>
<tr>
<th></th>
<th>Type 1/Type 2 Diabetes</th>
<th>CFRD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Calories</td>
<td>Calculated for maintenance, growth, or reduction diets</td>
<td>120-150% RDA</td>
</tr>
<tr>
<td>Carbohydrate</td>
<td>Individuallyized</td>
<td>Individuallyized</td>
</tr>
<tr>
<td>Fat</td>
<td>Individuallyized; often &lt;30% of total calories; &lt;10% saturated fat; ≤10% of calories from polyunsaturated fat</td>
<td>40% of calories; no restriction on type of fat</td>
</tr>
<tr>
<td>Protein</td>
<td>10-20% of total calories; reduction to 8.8 g/kg with nephropathy</td>
<td>10-20% total calories; no reduction with nephropathy*</td>
</tr>
<tr>
<td>Sodium</td>
<td>&lt;2,400 mg/day</td>
<td>&gt;4,000 mg/day</td>
</tr>
<tr>
<td>Vitamins/minerals</td>
<td>No supplementation unless deficiency noted</td>
<td>Routine supplementation of vitamins A, D, E, K, and multivitamin</td>
</tr>
</tbody>
</table>

*This is the recommendation of the consensus conference. In practice, a patient with severe nephropathy would require protein restrictions to prevent azotemia.

CFRD, cystic fibrosis-related diabetes; MNT, medical nutrition therapy; RDA, recommended dietary allowance.
**Future of Cystic Fibrosis**

- Today, advances in research and medical treatments have further enhanced and extended the lives for children and adults with CF.
  - Kalydeco
  - Lumacaftor
  - New Pancreatic Enzymes

**Questions?**

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**Thank You!!**

Did you know?

Cystic Fibrosis is sometimes called “65 Roses.” The nickname came from a little boy who overheard his mom talking about the condition on the phone. He thought that every time his mom said “Cystic Fibrosis,” she was talking about 65 roses.

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**References**

- "An Update on the Screening, Diagnosis, Management and Treatment of Vitamin D Deficiency in Individuals with Cystic Fibrosis: Evidence-Based Recommendations from the Cystic Fibrosis Foundation,” J Clin Endocrinol Metab. 2012;97(3):889-903
- "Clinical Care Guidelines for Cystic Fibrosis-Related Diabetes,” Diabetes Care. 2010 Dec;33(12):2697-2708