Meeting the Nutritional Challenges of Infants and Children Who Have Cystic Fibrosis

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Objectives

By the end of the session the participant will be able to:

• Describe two vitamins that can be deficient in CF

• Summarize Cystic Fibrosis Foundation (CFF) nutrition evidence-based guidelines

• Understand the role of optimal nutrition in the health of children who have CF

What is CF?

• CF is caused by abnormalities in cystic fibrosis transmembrane conductance regulator (CFTR) protein

• CFTR changes the function of mucosal epithelial cells
Normal CFTR protein serves as a channel that moves chloride and other ions through the cell membrane.

Symptoms of CF involve many organs.
CF Genetics

- > 1800 known CFTR mutations

- Mutations may be categorized as:
  - CF disease causing
  - Mutations with no clinical consequence
  - Mutations with uncertain/unknown clinical relevance

CF Genetics

- CF results from inheriting genetic mutations in the coding region for CFTR on chromosome 7

- Autosomal recessive inheritance
  - (need 2 mutations)
Before

Maximizing Growth and Nutrition in Patients With Cystic Fibrosis

Now
Newborn Screening for CF

- Is available in all states

Link Between Nutrition and Survival
Why is nutrition important?

- Weight (BMI)
- Height
- Lung Volume
- Survival

Median CDC Weigh Percentile vs. Age by Birth Cohort

Figure 28: Median CDC Weight Percentile vs. Age by Birth Cohort
Optimal nutrition status and weight early in life result in better pulmonary function later in life


- The strongest predictor of pulmonary function at age 6 was catch-up growth by age 2.
Topics

- Enzymes
- Energy
- Weight
- Vitamins
- Minerals

Pancreatic Function
Indirect Assessment of Pancreatic Function

- Coefficient of fat absorption (fecal fat study)
  - Not pancreatic specific

- Fecal elastase
  - Small amount of stool
  - Easy to do
  - Do not use watery stool sample

Supplemental pancreatic enzyme replacement therapy (PERT): Infants

- Start PERT in all infants with:
  - Two CFTR mutations associated with pancreatic insufficiency (PI)
  - A fecal elastase <200µg/g or CFA <85% or other objective evidence of PI
  - Unequivocal signs or symptoms of malabsorption, while awaiting confirmatory test results
    - Poor weight gain, abnormal stools, voracious appetite

Supplemental pancreatic enzyme replacement therapy (PERT): Infants

- Do not start PERT in infants with one or two CFTR mutations associated with pancreatic sufficiency unless:
  - An objective test of pancreatic function indicates fat malabsorption, or
  - The presence of unequivocal signs or symptoms of malabsorption, while awaiting confirmatory test results

Enzyme dosing: Infants

- Initiate at a dose of 2500 lipase units at each feeding, assuming feeding is 120 ml
- Adjust dose upward to a dose of not greater than 2500 lipase units per kg body weight per feeding
- Maximum daily dose of 10000 lipase unit per kg per day
Enzyme dosing: Children

- 500 to 2,500 lipase units per kg body weight per meal
- Snack dose is usually half the meal dose
- No more than 10,000 lipase units per kg per day
- <4,000 lipase units per gram of fat

Stallings, et al. 2008

Enzyme Products

- Creon (Abbott Labs)
  - 3, 6, 12, 24
- Pancreaze (Johnson & Johnson)
  - 4, 10, 16, 21
- Zenpep (Aptalis)
  - 3, 5, 10, 15, 20, 25
- Ultresa (Aptalis)
  - 13+, 20+, 23
- Pertzye (Digestive Care)
  - 8, 16
- Viokace (Aptalis)
  - Nonenteric-coated tablet
  - 8, 16
Method of Administering PERT to Infants

- Open capsule(s)
- Place dose of beads on a very small amount of applesauce
- Give enzymes to infant
- Feed bottle or breast milk

Calorie Recommendations

- 110% to 200% of requirements for the healthy population of the same age and gender

Stallings, et al. 2008
Energy

- **Increased Expenditure or Metabolic Issues**
  - Malabsorption
  - Cough
  - Inflammation
  - Work of breathing
  - Salt depletion
  - Increased activity

- **Appetite**
  - Iron deficiency
  - Zinc deficiency
  - GER
  - Suck/swallow challenges
  - Medications

- **Poor Absorption**
  - Enzyme Issues
  - Need for acid blockers
  - Suck/swallow challenges
  - Other GI conditions

- **Socioeconomic/Educational/Behavioral Issues**
  - Inadequate feeding knowledge
  - Maladaptive feeding behavior
  - Financial issues
  - Maladaptive family function


Type of Feeding (Infants)

- **Human milk**

- If fed formula, standard infant formulas, as opposed to hydrolyzed protein formulas
  - Hydrolyzed protein formula may be indicated for an infant who has had bowel surgery

- Calorie-dense feedings used if weight loss or inadequate weight gain is identified

Infant Feeding Studies


Diet for Children

- High calorie; high fat
  - Booster meals and snacks with behavior modification
  - Oral supplements
  - Tube-feeding
  - TPN
- Salt

- Case/control study: Matched for age, gender, pancreatic status, BMI%, and as available FEV1%.

- Supplementation to equal 50% of estimated needs based on CFF recommendations

- Supplements: 18 whole protein formula, 1 partially hydrolyzed formula, 1 elemental

- Results: Study suggests that those patients who receive GTs are more likely to achieve BMI ≥50% when compared to controls.

Weight Recommendations
FEV1 percent predicted is positively correlated with BMI percentiles for patients 6 to 20 years of age (correlation > 0.95, p < 0.0001).

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FEV1 percent predicted is positively correlated with BMI for patients 20 to 40 years of age (correlation = 0.92, p < 0.0001).
Weight recommendations

- Infants diagnosed before 2 yrs of age
  - Goal: weight-for-length of \textgreater{}50\textsuperscript{th} percentile by 2 yrs of age, based on CDC growth curve

- Children 2 to 20 yrs of age
  - Goal: BMI \textgreater{} 50\textsuperscript{th} percentile

- Adults
  - Goal: BMI
  - 22 kg/m\textsuperscript{2} for females
  - 23 kg/m\textsuperscript{2} for males


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Vitamins and CF

- Fat-soluble vitamins A, D, E, K

- Supplements need to be taken with fat containing food and enzymes
Vitamin A

- Combination of retinol and beta carotene
- Needed for:
  - Cellular integrity, growth, immune function, vision
  - Can be low with illness or zinc deficiency
- Avoid excess intake

Vitamin E

- Is an antioxidant
- Required for normal development
- Ataxia has been seen in CF patients
- Infants with lower serum vitamin E levels exhibited lower IQ points
Vitamin K

- Is needed for bone formation and coagulation.

- Persons with CF are at risk for deficiency due to:
  - use of antibiotics
  - bile salt deficiency
  - maldigestion with malabsorption
  - bowel resection
  - liver disease

- PT vs PIVKA

Vitamin D

- Bone health and other functions

- New CFF evidence-based report
### CFF Guidelines

<table>
<thead>
<tr>
<th>Routine Screening</th>
<th>25OHD (not 1,25OH2D or alk phos, osteocalcin, PTH, etc)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Timing</td>
<td>At annual bloodwork</td>
</tr>
<tr>
<td></td>
<td>Post dose adjustment</td>
</tr>
<tr>
<td></td>
<td>Ideally end of winter</td>
</tr>
<tr>
<td></td>
<td>3 months on new dose</td>
</tr>
<tr>
<td>25OHD Target</td>
<td>&gt;30 ng/mL</td>
</tr>
<tr>
<td>Vitamin D</td>
<td>Cholecalciferol</td>
</tr>
<tr>
<td>Formulation</td>
<td>Ergocalciferol</td>
</tr>
<tr>
<td>Frequency</td>
<td>Daily or weekly</td>
</tr>
</tbody>
</table>

**Tangpricha, et al. 2012**

<table>
<thead>
<tr>
<th><strong>Age</strong></th>
<th><strong>Routine Dosing with CF-specific vitamins (IU)</strong></th>
<th><strong>Step 1: Dose Increases (IU)</strong></th>
<th><strong>Step 2: Dose titration max (IU)</strong></th>
<th><strong>Step 3:</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth to 12 mo</td>
<td>400 - 500</td>
<td>800 - 1000</td>
<td>Not &gt; 2000</td>
<td>Refer &lt;10ng/mL</td>
</tr>
<tr>
<td>&gt;12 mo - 10 yr</td>
<td>800 - 1000</td>
<td>1600 - 3000</td>
<td>Not &gt; 4000</td>
<td>Refer</td>
</tr>
<tr>
<td>&gt;10 – 18 yr</td>
<td>800 - 2000</td>
<td>1600 - 6000</td>
<td>Not &gt; 10000</td>
<td>Refer</td>
</tr>
<tr>
<td>&gt;18 yr</td>
<td>800 - 2000</td>
<td>1600 - 6000</td>
<td>Not &gt; 10000</td>
<td>Refer</td>
</tr>
</tbody>
</table>
Vitamin Supplementation

- Prescribe CF-specific multivitamins designed to provide recommended levels of vitamins A, D, E, and K for patients with CF

- Measure blood levels of fat-soluble vitamins:
  - For infants approximately two months after starting vitamin supplementation and annually thereafter unless changing the vitamin dose
  - For newly diagnosed children: at the time of diagnosis and annually thereafter or 3 months following a change in vitamin prescription

- Measure more frequently if values are abnormal


CF-Specific Vitamin Supplements

- SourceCF
- Vitamax
- AquADEXK
Salt Supplementation for Term Infants

- Supplement with 1/8 teaspoon table salt per day starting at diagnosis
- Increase to ¼ teaspoon per day at six months of age


Salt Supplementation: Children

- High salt diet
- Routine use of foods high in salt
- Close attention in spring and summer

Borowitz D, et al. 2002
Fluoride and Zinc

- Supplement fluoride if water supply contains < 0.3 ppm. Refer to baby’s pediatrician.

- Trial of zinc supplementation with inadequate growth despite adequate caloric intake and optimal PERT or low serum retinol with adequate retinol intake.

- 1 mg elemental zinc/kg/day in divided daily doses for six months. Not to exceed 25 mg/day.

Iron and Calcium

- CF-specific vitamins do not contain iron.

- Provide supplemental iron if patient has iron deficiency anemia.

- Calcium from diet or if needed supplements.
Feeding Behavior

• Positive feeding behaviors is encouraged

• For infants and children with growth deficits intensive treatment with behavioral intervention in conjunction with nutrition counseling be used to promote weight gain

Stallings V, et al. 2008

Cystic Fibrosis Foundation Evidence-based guidelines for management of infants with Cystic Fibrosis

Evidence-based practice recommendations for nutrition-related management of children and adults with Cystic Fibrosis and pancreatic insufficiency: Results of a systematic review


An update on screening, diagnosis, management and treatment of vitamin D deficiency in individuals with Cystic Fibrosis: Evidence-based recommendations from the Cystic Fibrosis Foundation.

Summary

- The guidelines offer evidenced-based recommendations for the RD to apply for:
  - The care of infants diagnosed through newborn screening
  - The management of energy, enzyme, growth and vitamin D of individuals who have CF

Summary

- RD plays a pivotal role in the care of persons diagnosed with CF
- Optimal nutrition early in life equals improved health later in life
- Optimal nutrition throughout life equals improved health
Thank you!

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