Cystic Fibrosis and Nutrition

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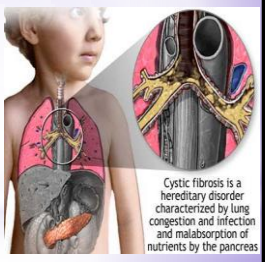
Learning Objectives

- 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.

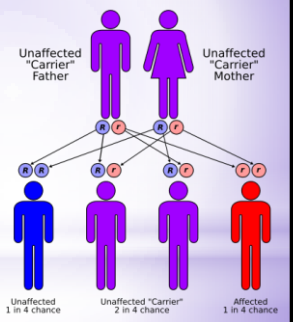


Statistics

- 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.

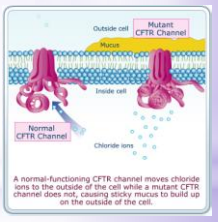
Diagnosing Cystic Fibrosis

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



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_3, E & K
    - CF Vitamins:
      - AquaDEK – **Liquid,** Chewable & **Gel Capsule**
      - SourceCF – Liquid, Chewable & Gel Capsule
      - Vitamax – 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

Age	Liquid	Chewable	Soft-Gel
0-12 months	1 ml (0.5 ml BID)		
1-3 years old	2 ml (1 ml BID)		
4-8 years old		1 tablet	
9-18 years old			2 Capsules (1 Capsule BID)
>18 years old			2 Capsules (1 Capsule BID)

- Additional supplementation:
  - Vitamin A: 8,000 – 10,000 IU once daily
  - Vitamin D: 10,000 – 50,000 IU weekly
- Vitamins with meals and enzymes!

Signs of Maldigestion 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

Enzyme	Strengths (USP Units of Lipase)
Creon 24,12,6 &3	24000,12000,6000 & 3000
Zenpep 25,20,15,10,5 & 3	25000, 20000, 15000, 10000, 5000 & 3000
Ultresa 23,20, & 13	23000, 20700 & 13800
Viokase 16 & 8	16000 & 8000
Pancreaze 21,16,10 & 4	21000, 16800, 10500 & 4200
Pertzye 16 & 8	16000 & 8000

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.

Calculating Pancreatic Enzymes

- 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

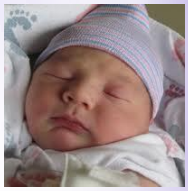
Calculating Pancreatic Enzymes

- 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

	Red Nutritional Failure	Yellow Nutritional Risk	Green Nutritional Health
Infants (0-23 months)	≤10% ht-wt	11-49% ht-wt	≥ 50% ht-wt
Children (2-20 yrs. of age)	≤10% BMI for Age	11-49% BMI for Age	≥ 50% BMI for Age
Adults (20 yrs. of age and older)	≤ 18 kg/m^2 BMI	18-22 kg/m^2 BMI Males 18-21 kg/m^2 BMI Females	23 kg/m^2 BMI Males 22 kg/m^2 BMI Females

Comorbidities Related to Cystic fibrosis

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

Cystic Fibrosis Related Diabetes (CFRD)

- Cystic Fibrosis-Related Diabetes (CFRD) is a unique type of diabetes
  - It is NOT the same as Type I or Type II 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

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

	Type 1/Type 2 Diabetes	CFRD
Calories	Calculated for maintenance, growth, or reduction diets	120–150% RDA
Carbohydrate	Individualized	Individualized
Fat	Individualized; often <30% of total calories, <10% saturated fat, ≤10% of calories from polyunsaturated fat	40% of calories; no restriction on type of fat
Protein	10–20% of total calories; reduction to 0.8 g/kg with nephropathy	10–20% total calories; no reduction with nephropathy*
Sodium	<2,400 mg/day	>4,000 mg/day
Vitamins/minerals	No supplementation unless deficiency noted	Routine supplementation of vitamins A, D, E, K, and multivitamin

*This is the recommendation of the consensus conference.[8] In practice, a patient with severe nephropathy would require protein restriction to prevent azotemia.

CFRD, cystic fibrosis-related diabetes; MNT, medical nutrition therapy; RDA, recommended dietary allowance

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

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

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

greatstrides
TAKING STEPS TO CURE CYSTIC FIBROSIS

CYSTIC FIBROSIS FOUNDATION
ADDING TOMORROWS

Questions?

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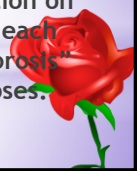
Thank You!!

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 each time his mom said "Cystic Fibrosis" she was talking about 65 roses.



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