SCHOOL OF PUBLIC HEALTH
UNIVERSITY of WASHINGTON

Medical Nutrition Therapy for Pediatric Cystic Fibrosis Patients

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Definition of Cystic Fibrosis

- Autosomal recessive genetic disorder causing systemic dysfunctions of exocrine glands.
- Production of thick mucus throughout the body
- Clogged airways
- Interfere with pancreatic enzyme production
- Related conditions: CF-related diabetes mellitus, liver and gall bladder disease

Nutrition Implication

Increased Energy Needs
- 130%-150% recommended caloric intake

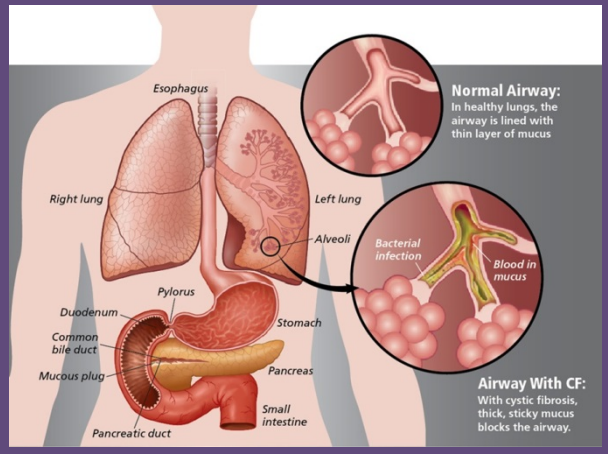
Increased Protein Needs
- 200% recommended dietary allowance

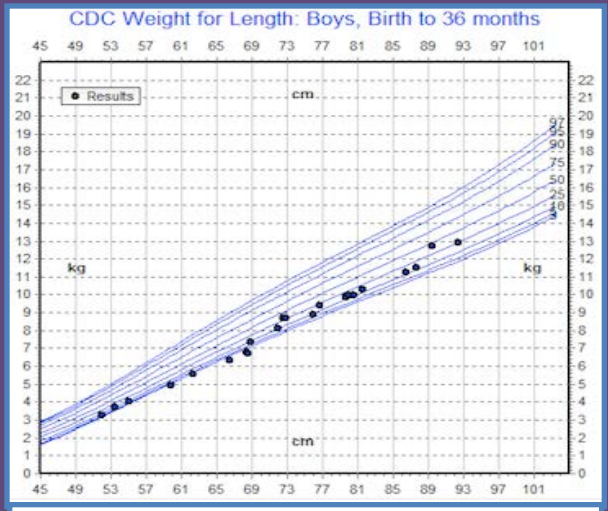
Pancreatic Insufficiency
- Fat and fat soluble vitamin malabsorption
- Pancreatic enzyme replacement therapy

Bone Health
- Vitamin D supplementation

GI symptoms
- Cramping might benefit from high fiber diet

Acknowledgement
Site: Seattle Children's Hospital
Advisor: Susan Casey RD, CD.
Figure credited to Discover magazine





Growth trajectory of Little J. His weight for length raised from 5th percentile to 14th percentile after appropriate management of Cystic Fibrosis.

Nutrition Goals

- Higher weight relates to better lung function
- Weight for Length at 50th percentile for 2yrs. And under
- BMI percentile at or above 50th percentile for 2-20 yrs

Case Study

History Little J 28 months old boy with Cystic Fibrosis

Other diagnosis
MRSA Positive and Pancreatic Insufficiency

Psychosocial
Lives with both parents. Mom is fearful about Little J getting a feeding tube. She pushed his feeding quite a lot, by using heavy whipping cream mixed with all his foods and feeding him 9 meals per day. Little J experienced some GI symptoms related to fat malabsorption, which dissolved after mom stopped feeding him excessive fat.

Anthropometric Assessment
Weight: 12.9 kg (36th percentile, weight of 27 months)
Height: 92.45cm (69th percentile, height of 33 months)
BMI: 15.09 (14th percentile)
Ideal Body weight: 14.25 kg, Percent of Ideal Body Weight: 91%
Gained 200 grams in the past 3 months

Diet and Medication Assessment
- Mom overcompensated previous overfeeding by restricting snacks to non-fat products such as popsicles. Not sure whether enzymes worked after 45 minutes
- Creon 6000, 4 per meal and 3 per snack
- Multivitamin: AquADEK

Estimated Nutrient Needs
Calories (1.3 X RDA) = 1889 kcal
Protein (2 X RDA) = 34 g
Fluid = 1145 ml

Nutrition Diagnosis
Inadequate energy intake (NI-1.4) related to cystic fibrosis as evidenced by BMI below 20 percentile for age.

Nutrition Intervention
- Education on enzyme use (effective for 2 hours)
- Revise food and nutrient delivery: stop restricting snacks to non-fat items
- Keep identifying and feeding high calorie, high protein food items
- Keep current pancreatic enzyme intake
- Keep supplementation of multivitamin

Monitor and Evaluation
- Growth: Appropriate weight gain and height growth. Goal BMI at 50th percentile.
- Diet intake: 3meals and 3 snacks taken with pancreatic enzymes. Make sure adequate caloric and protein intake
- Biochemical: Check vitamin D status regularly