CYSTIC FIBROSIS: NUTRITION INTERVENTIONS FOR THE ADULT POPULATION

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Overview

What is RDs role in cystic fibrosis?

1. Fat-Soluble Vitamins
2. Sodium
3. Pancreatic Enzyme Replacement Therapy (PERT)
4. Cystic Fibrosis Related Diabetes (CFRD)
5. Body Mass Index (BMI) and Nutrition Status
BMI in Adults with CF

- Accepted measure of nutritional status in people with CF. 1, 3
  - Women: BMI of 22
  - Men: BMI of 23

- Calculated at each visit

- Why is BMI used?
  - The BMI percentile was shown to be more sensitive to changes in pulmonary function than %IBW. 3
  - New research has shown that lean body mass was a greater predictor of nutritional status than BMI. 8
Case Study - Assessment

- 22 year old, male
- Admitted for CF exacerbation - Endobronchitis
- CF related diabetes
- Pancreatic insufficient - PERT
- 9% weight loss in one month
  - BMI=20.51 kg/m²
- Diet order: Regular, high kcal/high fat
Cystic Fibrosis-Related Diabetes (CFRD)

- Symptoms:
  - No symptoms (majority)
  - Polyuria or polydipsia
  - Unexplained difficulty gaining or maintaining weight despite interventions
  - Poor growth
  - Unexplained decline in pulmonary function
CFRD continued

- Most common comorbidity in CF
- Different pathophysiology than Type 1 or 2 DM
- Loss of islets $\rightarrow$ $\beta$-cell destruction $\rightarrow$ no insulin
- During illness, insulin sensitivity becomes significantly decreased
- Pulmonary deterioration has been correlated with severity of insulin insufficiency
CFRD – Diagnosis & Treatment

- A1c levels are often falsely low
- Oral glucose tolerance test
  - The North American CFRD Guidelines Committee determined accepted measure
- Treated with insulin
- Oral diabetes agents are less effective with CFRD
  - Only used in clinical research trials
CFRD Treatment continued...

- **Calories:** 1.2-1.5 times DRI for age.⁹
- **Carbohydrates:** Individualized to achieve glycemic control.⁹
- **Fat:** No restrictions.⁹
- **Protein:** 1.5-2.0 times DRI for age.⁹
- **Sodium:** High salt diet.⁹
- **Weight loss:** Not recommended, eat carbohydrates throughout the day, and choose nutrient dense beverages.⁹
- **Vitamins & minerals:** Multivitamin and fat soluble vitamins.⁹
Pancreatic insufficiency - Pancreatic ducts are blocked, preventing enzymes from properly digesting food. Insufficiency in digestion of protein, fat, and fat soluble vitamins, may lead to poor nutrition status. Fat soluble vitamins in micelle form have a better chance at absorption.²

Dosing based on lipase between 500 – 2500 units/kg, for children and adults, not to exceed 10,000 units lipase/kg per day.³

Maximum patient dose at 60 kg = 600,000 units/day

60 kg x 4,400 units/kg/d = 264,000 units per day
- 6 tabs 24,000 units/tab with meals 4 x day = 144,000 units
- 5 tabs 24,000 units/tab with snacks 4x day = 120,000 units
Nutrition Focused Physical Exam

- Screening tool to diagnose malnutrition
  - Fat loss
  - Muscle loss
- Vitamin and mineral deficiencies
- Nutrition status outside of BMI
Diagnosis

- Inadequate oral food/beverage intake related to poor appetite as evidenced by 9% patient reported weight loss in one month.

- Increased nutrient needs: calories/protein related to cystic fibrosis as evidenced by estimated needs greater than daily recommended intake (DRI) for standard age.
Individually Tailored Interventions

- Age
- Nutrition & Pancreatic Status
  - PERT / Diabetes / BMI
- Financial & Family Circumstance
- Religious & Cultural Dietary Beliefs
- Food Preferences
Intervention

- **Energy needs** 110 – 200%. \(^2, 3\)
  - Patient: 52-60 kcal/kg/day = 3088 – 3564 kcal/day

- **Protein needs** 1.5 g/kg. \(^2\)
  - Patient 1.2 – 1.5 grams/kg/day = 70 – 90 grams daily

  - RD added clear protein drinks at least TID
    - 250 kcal, 9 grams protein each
  
  - Protein milkshakes made with protein powder, ice cream, and half ’n’ half at least BID
    - 361 kcal, 24 grams protein each
  
  - Supplement total at least 1472 kcal & 75 grams of protein daily
Oral Nutrition Supplements

- Can help to prevent weight loss during acute respiratory exacerbations.\(^2\)
- Sodium supplements
- CF requires double the normal RDA for fat-soluble vitamins.\(^2\)
- Deficiency of fat soluble vitamins A, D, E, and K in pancreatic insufficient individuals.
Intervention continued

- Added vitamin A, and vitamin E
  - Patient was not on vitamin A, in hospital
  - Patient’s vitamin E level was low
    - Coordination of care, doctor & pharmacy
Patient Outcome

- Patient discharged about 2 weeks after admission.
- Gained back the weight lost before hospital admission.
Why is nutrition important?

- Nutrition is important in life expectancy outcomes
  - Oral nutrition supplements,
    - ADEKs
    - Sodium
  - PERT
  - Diabetes
  - BMI & Nutrition Status
The average life expectancy today is around 37 years.  

Babies born today with CF have a predicted life expectancy between 40 – 50 years.
Questions?


