Cystic Fibrosis: A Definition

CF is: “a disease that is characterized by abnormally thick mucus secretions from the epithelial surfaces of various organ systems” (Nelms, 2007)

Organ systems involved:

- The respiratory tract
- The GI tract
- The liver
- The reproductive system
- The sweat glands
COMMON SYMPTOMS

- Very salty-tasting skin
- Persistent coughing, at times with phlegm
- Frequent lung infections, such as pneumonia or bronchitis
- Wheezing or shortness of breath
- Poor growth/weight gain in spite of a good appetite
- Small, fleshy growths in the nose called nasal polyps
EPIDEMIOLOGY

○ CF is the most common **autosomal recessive** disease in the US *(Nelms, 2007)*
  - Affects approx. 30,000 children and adults
  - When two people, each with one defective gene, conceive:
    ○ 25% chance the child will have CF
    ○ 50% chance the child will be a CF gene carrier
    ○ 25% chance the child will not be a CF carrier

○ Most commonly diagnosed:
  - In children, by age 3 years
  - In Caucasians

○ Life expectancy is, on average, 37 years
Etiology

- The mutation of the cystic fibrosis transmembrane conductance regulator (CFTR) gene (Nelms, 2007)
  - Found on chromosome 7
  - Is a protein
PATHOPHYSIOLOGY

- CFTR protein responsible for the transport of:
  - Sugars, peptides, inorganic phosphates, chloride, metal cations across cell membranes
  - Chloride ions across cell membranes in the lungs, liver, pancreas, digestive tract, reproductive tract, skin

- In other words... the transport of salt and water in and out of the epithelial cells is disturbed
  - Results in a hyper-secretion of viscous mucus from the exocrine glands
  - Leads to obstruction of glands and ducts
**Pathophysiology**

- **Respiratory**
  - Chronic coughing and wheezing
  - Might begin during the first month of life
  - Respiratory failure and death is a result in 90% of pts
  - Clogged airway → infection → respiratory failure

- **Pancreatic**
  - 85-90% of pts
  - Bulky, foul smelling stool, abd. distention, poor growth
  - Insulin-dependent diabetes, approx. 10% of adults
PATHOPHYSIOLOGY

- Reproductive
  - Men become infertile
  - For women, pregnancy can ↑ signs/symptoms of CF

- Gastrointestinal
  - ↓ pancreatic enzymes  ➔ ↓ absorption of nutrients
  - Folded intestines (Intussusception) can lead to an obstruction
  - Blocked bile ducts ➔ liver problems
DIAGNOSTIC TESTS

- Sweat chloride test
- Blood test for the CFTR gene
- Pancreatic function test
- Less sensitive tests:
  - Sputum cultures
  - Spirometry
  - Chest radiographs
Nutrition Implications

- Respiratory:
  - Anorexia, ↑ Energy requirements, malnutrition

- Pancreatic:
  - Inadequate digestion, nutrient malabsorption (fat & fat sol. vits.)
  - under wt., growth failure, and delay in puberty
  - Osteoporosis and osteopenia

- GI: (due to pancreatic insufficiency)
  - ↓ absorption of pro, fat, fat sol. vitamins
  - Loss of bile and bile salts
NUTRITION IMPLICATIONS

FIG 1. Interdependent factors that contribute to progressive energy deficit in cystic fibrosis.
CASE STUDY

- **Client Name:** Lily Johnson
- **Age:** 14
- **Sex:** Female
- **Education:** Just completed 9th grade
- **Household members:** Mother age 41 (divorced), grandmother age 66 (widowed), half-brother age 5
- **Ethnic Background:** Caucasian
**CASE STUDY**

- **Chief complaint:**

  “I just got back from working at a camp for the past two weeks. I caught a cold, and it has just gotten worse. My regular treatments were not working, and my doctor says I probably have pneumonia.”
CASE STUDY: PATIENT HISTORY

- Onset of disease:
  - Lily was diagnosed at age 6 months with CF
  - Has had a rather uneventful disease course
  - Hospitalized several times with respiratory infection, otherwise has maintained her disease with outpatient therapy
  - Has yearly visits to the CF clinic at the Univ. hospital, and receives routine med. care from her local physician
CASE STUDY: PATIENT HISTORY

- **Medical treatment:**
  - High frequency chest compression vest, 1 hr x 2x/day

- **PMH:**
  - Last hospitalization over a year ago, successful first year of HS
  - She is very active: ballet and jazz, cross-country runner
    - Runs 3-5 mi, 5-6 times/week
    - Dance class 3x/week
Case Study: Medications

- Pancrease: to treat steatorrhea 2° pancreatic insufficiency
-Prevacid/Prilosec: antiulcer, antiGERD
- Humabid: expectorant
- MVI
- Proventil: bronchodilator
CASE STUDY: PHYSICAL EXAM

- **General Appearance:** 14 y/o thin female, flushed, no acute distress
- **Throat:** Pharynx reddened with postnasal drainage
- **Skin:** skin pale w/o rash
- **Chest/lungs:** Decreased breath sounds, percussion hyperresonant, rhonchi and rales present
Case Study: Nutrition Hx

- Previous Nutrition Therapy:
  - Nutrition info received at CF clinic
  - Meeting with dietitian
  - “Just recently started thinking about my diet. My family has really made most of those decisions. I know I need to know more about my diet, and I really want to make sure I stay healthy.”

- Food allergies/intolerance/aversions:
  - Will eat almost anything
  - Tries to avoid fried foods ➔ diarrhea
CASE STUDY: DIAGNOSIS

- **Acute pneumonia**
  - Confirmed by chest x-ray (CXR) and sputum culture

- **Hospital Course:**
  - IV antibiotics were initiated
  - Nutrition consult to assess current nutritional status and to ensure adequacy of current nutritional intake

- **Treatment Plan:**
  - Bed rest with a regular diet as tolerated
  - Lab: CBC, RPR, Chem 16: I &O every shift; routine vital signs
  - IVF D₅ @ 50ml/kg IV q6h
ASSESSMENT PARAMETERS

- Screening for malnutrition
  - Anthropometric indexes: weight-for-age, weight-for-height, or %IBW*, BMI percentiles (BMIp)
  - For adults: BMI
  - Use of both %IBW and BMIp in combination (Wiedemann, 2007)

- Nutrition/Behavior (Powers, 2005)
  - Number of meals eaten/day
  - Enzyme usage
  - Number of kcal consumed/day
  - High fat food choices
ASSESSMENT

- Psychological
  - Acceptance and adjustment to the disease (Casier, 2008)
- CF Related Diabetes (CFRD)
  - Fasting hyperglycemia (plasma glucose >7.0 mmol/L) on more than 2 separate occasions
  - Fasting hyperglycemia on one occasion and a random glucose level >11.1 mmol/L
CASE STUDY ASSESSMENT: ANTHROPOMETRICS

- Anthropometric Data:
  - Ht 5’5”
  - Wt 102 lbs
  - UBW 110-115 lbs (3 mo ago)
- Wt/age = 42$^{nd}$ percentile
- Stature/age = 75$^{th}$ percentile
- BMI/age = 18$^{th}$ percentile
CASE STUDY ASSESSMENT: NUTRITION HX

- General:
  - Appetite fine until last few days
  - Never really know how much Pancrease to take
  - Never drinks milk
  - Likes fruit and veg. but doesn’t eat them much
  - Rarely eats breakfast

- Food purchase/preparation:
  - Self, mother, grandmother

- Vitamin/mineral intake:
  - Tries to remember MVI, but not taken every day
**CASE STUDY ASSESSMENT: NUTRITION HX**

- **Usual dietary intake:**
  - **AM:** rarely eats
  - **Lunch:** 3 tbls peanut butter or 2oz ham w/ 2 oz swiss cheese sandwich, 2-3 oz chips, 1pc fruit, water
  - **Dinner:** 5-6 oz pro (grilled or baked), 1-2C raw veg on lettuce, ¼C ranch dressing, 1C starch w/ 1-2 tbsp margarine, water

- **24-hr Recall**
  - **AM:** Nothing
  - **Lunch:** 2 oz hotdog on a bun, 1 ½C macaroni & cheese (Kraft, w/ 2% milk)
  - **Dinner:** 5oz Salisbury steak, 1/4 C gravy, few bites green beans, 1 roll w/ 2 tbls margarine, approx 2C grape juice
### Case Study Assessment: Lab Work

<table>
<thead>
<tr>
<th>Lab</th>
<th>Normal</th>
<th>Actual</th>
<th>Indication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Transferrin</td>
<td>250-380</td>
<td>219 (L)</td>
<td>Chronic illness/inflammation/meds</td>
</tr>
<tr>
<td>Magnesium</td>
<td>1.8-3</td>
<td>1.6 (L)</td>
<td>Malabsorption/malnutrition</td>
</tr>
<tr>
<td>HDL</td>
<td>&gt;55</td>
<td>55 (L)</td>
<td></td>
</tr>
<tr>
<td>HbA1c</td>
<td>3.9-5.2</td>
<td>6.3 (H)</td>
<td>Pancreatic insufficiency</td>
</tr>
<tr>
<td>WBC</td>
<td>4.8-11.8</td>
<td>13 (H)</td>
<td>pneumonia</td>
</tr>
<tr>
<td>Hgb</td>
<td>12-15</td>
<td>11.5(L)</td>
<td>Iron def. anemia/meds</td>
</tr>
<tr>
<td>Hct</td>
<td>37-47</td>
<td>33 (L)</td>
<td>Iron def. anemia/meds</td>
</tr>
<tr>
<td>Ferritin</td>
<td>20-120</td>
<td>19 (L)</td>
<td>Iron def. anemia/meds</td>
</tr>
</tbody>
</table>
# Case Study: Medication Implications

<table>
<thead>
<tr>
<th>Medication</th>
<th>Nutrition Implication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pancrease</td>
<td>↓ Fe and folate absorption</td>
</tr>
<tr>
<td>Prilosec/Prevacid</td>
<td>May ↓ Fe abs, ↓ B&lt;sub&gt;12&lt;/sub&gt; abs, may ↓ Ca abs</td>
</tr>
<tr>
<td>Humabid</td>
<td>May cause N/V, diarrhea, GI distress</td>
</tr>
<tr>
<td>Proventil</td>
<td>↑ Appetite, anorexia; ↓ vit K, Hgb, Hct</td>
</tr>
</tbody>
</table>
CASE STUDY: MACRONUTRIENT REQ’S.

- **Energy Needs:**
  - DRI for 14y/o = 2400kcal/day
  - For CF, aim for >100% RDA

- **Fat:** 35-40% kcals
  - 840-960 kcal from fat/day
  - Or 93-107g fat per day

- **CHO:** 40-50% kcals
  - 960-1200 kcal from CHO/day
  - Or 240-300g CHO per day

- **Pro:** 15-20% kcals
  - 360-480 kcal from protein/day
  - Or 90-120g pro per day
CASE STUDY: MICRONUTRIENT REQS.

- Calcium
  - Need 1300 mg/day
  - Getting: 532 mg
- Magnesium
  - Need 360 mg/day
  - Getting: 235 mg
- Potassium
  - Need 4700 mg/day
  - Getting: 3537 mg
- Fat soluble vitamins (A, E, D, K)
**DIAGNOSIS**

- Inadequate energy intake (NI-1.4) related to poor appetite as evidenced by analysis of dietary recall (usual intake is 86% of calculated needs)

- Involuntary weight loss (NC-3.2) related to increased physical activity, poor appetite, and steatorrhea as evidenced by 11% wt. loss p 3 mos.

- Food and nutrition-related knowledge deficit (NB-1.1) related to lack of education regarding pancrease dosage and age-related readiness to take more responsibility for self care as evidenced by pt. report of not knowing how much pancrease to take, and steatorrhea in spite of taking meds.
INTERVENTION

- Treatment plans that affect nutritional outcomes in CF
  - Caloric intake, nutrient absorption, CFRD
- Behavior modification
- Nutritional Supplement intake
  - Oral (MVI)
  - Possible TF (gastrostomy tube), short- or long-term supplemental enteral or parenteral nutrition
    - However, no conclusive information is currently available for suppl. enteral/parenteral tube feeding for CF (Conway, 2010)
CASE STUDY: INTERVENTION

#1: Meals and Snacks (ND-1): General/Healthful Diet

- **Outcome goals:**
  - Increase daily energy intake (total kcal, pro, and CHO)
  - Modify the distribution, type, or amount of food and nutrients within meals

- **Action goals:**
  - Include breakfast in her daily meal pattern
  - Include 1-2 snacks per day in her diet
  - Be able to balance her meals with CHO, pro and fat
  - Include \( \frac{1}{2} \) to 1 cup fruit or veg. with each meal
CASE STUDY: INTERVENTION

#2: Comprehensive Nutrition Education (E-2)

- Outcome goals:
  - ≥2400 kcal per day, with 35-40% of kcals from fat
    - Action: Completion of 24-hr recall, or food journal
  - Demonstrates understanding appropriate Pancrease dosage for a given meal
  - Pt. weight management- increase weight to a proper maintenance weight
    - To be at 50% BMI-for-age: need to gain 12 lbs (to 114 lbs)
CASE STUDY: INTERVENTION

#3: Vitamin and Mineral Supplement (ND-3.2)

- **Outcome goals:**
  - Improved nutrition related lab values
    - Transferrin, Mg, Hgb, Hct

- **Action goals:**
  - Improve adherence to multivitamin intake
Intervention #1: Meals and Snacks (ND-1)

Intervention #2: Comprehensive Nutrition Education (E-2)

Intervention #3: Vitamin and Mineral Supplement (ND-3.2)
**Monitor and Evaluate**

- Caloric intake
- Weight
- Adherence to nutrition-related guidelines
  - MVI
- Labs:
  - Transferrin, Mg, HDL, Hgb, Hct, Ferritin, HbA1c, Ca^{2+}
**Case Study: Monitor and Evaluate**

- **Follow-up:**
  - Outpatient dietitian: meet PRN
    - Review diet recall
    - Review new/changed behavior
      - MVI included in diet?
      - Eating breakfast and snacks?
    - Food intake
      - Including more kcal, complex CHO
REFERENCES


REFERENCES, CONT.


- Pronsky Z, Crowe J. *Food Medication Interactions, 16th Edition* Birchrunville, Pa 2010

