



# **CYSTIC FIBROSIS**

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## 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

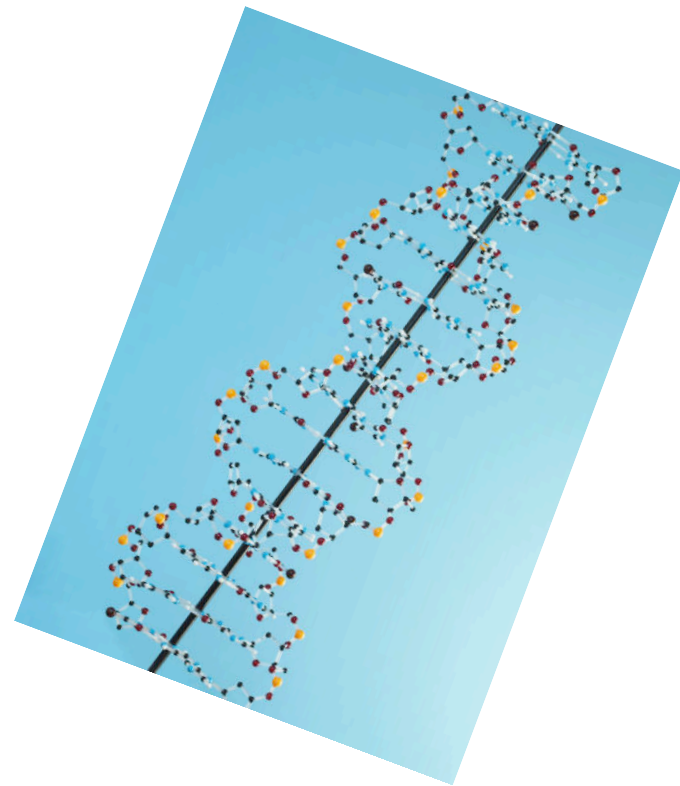
- Very salty-tasting skin
- Persistent coughing, at times with phlegm
- Frequent lung infections, such as pneumonia or bronchitis
- Wheezing or shortness of breath
- Poor growth/weight gain in spite of a good appetite
- Small, fleshy growths in the nose called nasal polyps

# 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

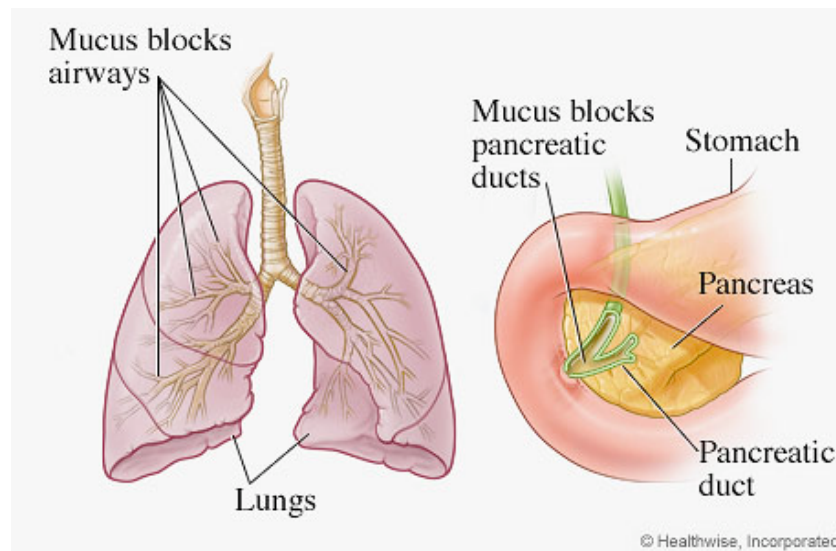
- The mutation of the cystic fibrosis transmembrane conductance regulator (CFTR) gene (Nelms, 2007)
  - Found on chromosome 7
  - Is a protein



# 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

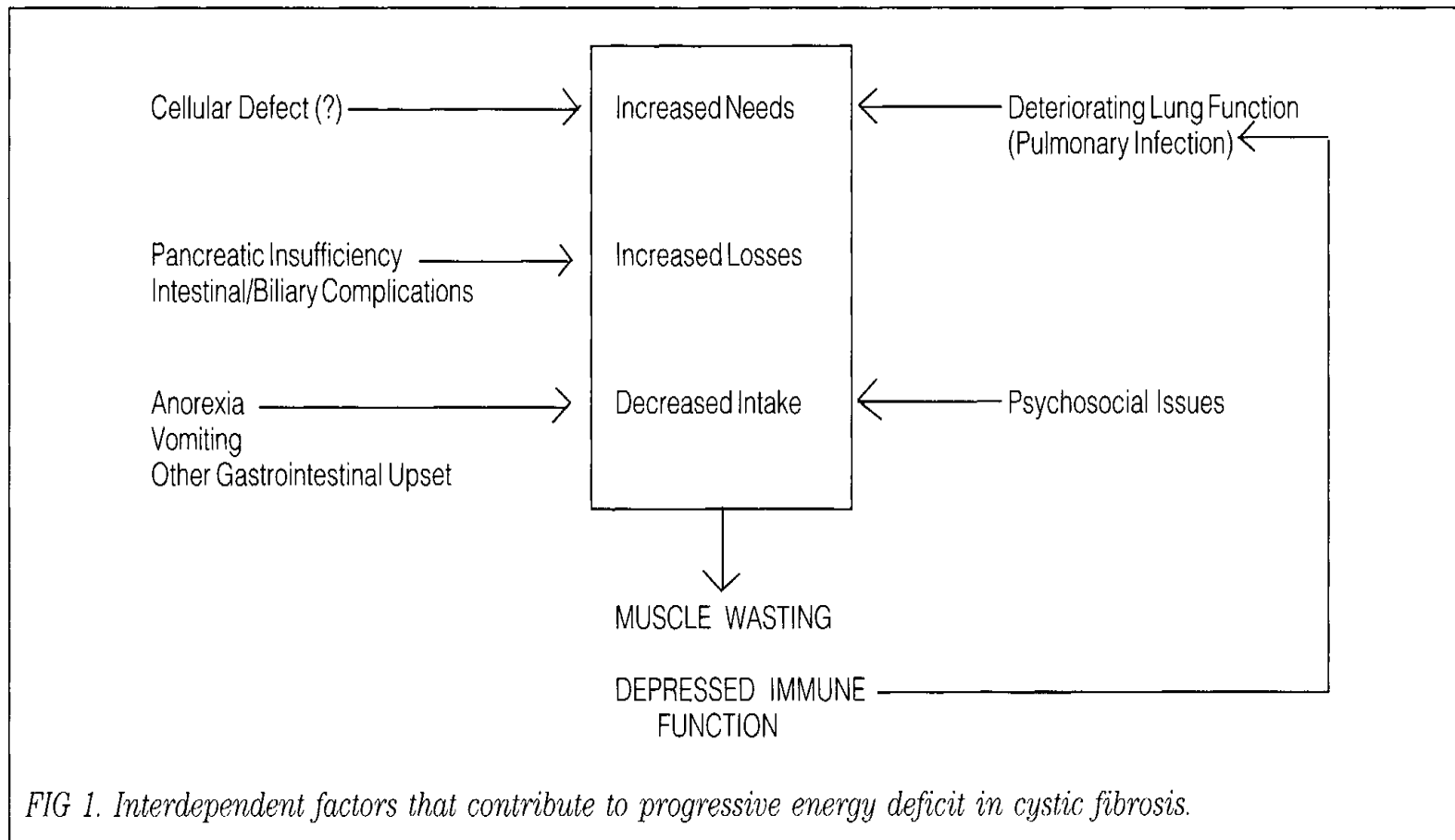


FIG 1. Interdependent factors that contribute to progressive energy deficit in cystic fibrosis.

## CASE STUDY

- Client Name: Lily Johnson
- Age: 14
- Sex: Female
- Education: Just completed 9<sup>th</sup> 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 D<sub>5</sub> @ 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<sup>nd</sup> percentile
- Stature/age = 75<sup>th</sup> percentile
- BMI/age = 18<sup>th</sup> 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

<u>Lab</u>	<u>Normal</u>	<u>Actual</u>	<u>Indication</u>
Transferrin	250-380	219 (L)	Chronic illness/ inflammation/meds
Magnesium	1.8-3	1.6 (L)	Malabsorption/ malnutrition
HDL	➤55	55 (L)	
HbA1c	3.9-5.2	6.3 (H)	Pancreatic insufficiency
WBC	4.8-11.8	13 (H)	pneumonia
Hgb	12-15	11.5(L)	Iron def. anemia/meds
Hct	37-47	33 (L)	Iron def. anemia/meds
Ferritin	20-120	19 (L)	Iron def. anemia/meds

## CASE STUDY: MEDICATION IMPLICATIONS

<u>Medication</u>	<u>Nutrition Implication</u>
Pancrease	↓ Fe and folate absorption
Prilosec/Prevacid	May ↓ Fe abs, ↓ B <sub>12</sub> abs, may ↓ Ca abs
Humabid	May cause N/V, diarrhea, GI distress
Proventil	↑ Appetite, anorexia; ↓ vit K, Hgb, Hct

## 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 ½ to 1 cup fruit or veg. with each meal



## CASE STUDY: INTERVENTION

- #2: Comprehensive Nutrition Education (E-2)
  - Outcome goals:
    - $\geq 2400$  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

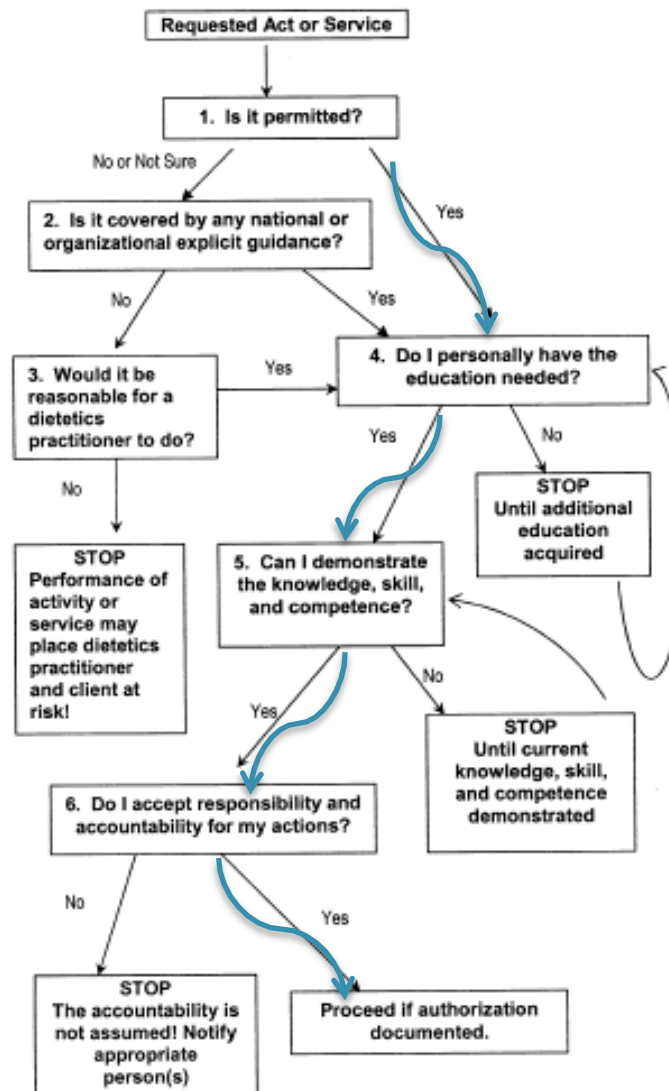
- #3: Vitamin and Mineral Supplement (ND-3.2)
  - Outcome goals:
    - Improved nutrition related lab values
      - Transferrin, Mg, Hgb, Hct
  - Action goals:
    - Improve adherence to multivitamin intake

# SCOPE OF PRACTICE

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<sup>2+</sup>

# 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

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