The Client with Cystic Fibrosis

Study Guide
Use Hockenberry and Lewis textbooks to complete this study guide.

1. Define Cystic Fibrosis (Mucoviscidosis)
   An old name (but one that has prevailed in France and some other nations) for cystic fibrosis (CF), one of the most common and serious of all genetic (inherited) diseases. The CF gene is carried by 1/20 persons (in Caucasian populations) and 1 in 400 couples is at risk for having children with CF. CF is characterized by the production of abnormal secretions leading to mucous build-up, which can impair the pancreas (and, secondarily, the intestine). CF mucous build-up in lungs can impair respiration. Without treatment, CF results in death for 95% of children before age 5. Early diagnosis of CF is of great importance. Early and continuing treatment of CF is valuable.
   - Autosomal recessive multisystem disease (inherited) of exocrine glands, lungs, pancreas, sweat glands and sex glands
   - Most common genetic disorder in white children
   - 2004 survival rate was 35 years
   - Most didn’t live to adolescence when she started it was 16-20 now up to 40
   - 2 recessive genes, I from mom and 1 from dad and there are carriers
   - If you have 1 child with cystic, it doesn’t take away the 1 in 4 chance
   - They can isolate the gene and do a blood study to check for the problem
   - 1 in 20 americans are possible carriers, symptom free
   - Screen infant
   - Screen in amniocentesis
   - Acid base risk – Respiratory acidosis, they can’t exchange the gases easily, always compensating

2. Describe the cause and the pathophysiology.
   The protein created by this gene is anchored to the outer membrane of cells in the sweat glands, lungs, pancreas, and other affected organs. The protein spans this membrane and acts as a channel connecting the inner part of the cell (cytoplasm) to the surrounding fluid. This channel is primarily responsible for controlling the movement of halogen from inside to outside of the cell; however, in the sweat ducts it facilitates the movement of chloride from the sweat into the cytoplasm. When the CFTR protein does not work, chloride and thiocyanate are trapped inside the cells in the airway and outside in the skin. Then hypothiocyanite, OSCN, cannot be produce by immune defence system. Because chloride is negatively charged, positively charged cations cross into the cell because they are affected by the electrical attraction of the chloride ions. Sodium is the most common ion in the extracellular space and the combination of sodium and chloride creates the salt, which is lost in high amounts in the sweat of individuals with CF. This lost salt forms the basis for the sweat test.
   - Mutation on 7th chromosome, a protein on there is not normal
   - The mutant intereferes with cystic fibrosis transregulator
     - Mutations are in the linings of the ducts
     - Failure to thrive

3. Explain the effects of cystic fibrosis on each of the body systems (organs):
   a. Respiratory tract (bronchi)
Lung disease results from clogging the airways due to mucosa build-up and resulting inflammation. Inflammation and infection will cause injury and structural changes to the lungs, leading to a variety of symptoms. In the early stages, incessant coughing, copious phlegm production, and decreased ability to exercise are common. Many of these symptoms occur when bacteria that normally inhabit the thick mucus grow out of control and cause pneumonia. In later stages of CF, changes in the architecture of the lung further exacerbate chronic difficulties in breathing. Other symptoms include coughing up blood (hemoptysis), changes in the major airways in the lungs (bronchiectasis), high blood pressure in the lung (pulmonary hypertension), heart failure, difficulties getting enough oxygen to the body (hypoxia), and respiratory failure requiring support with breathing masks such as bilevel positive airway pressure machines or ventilators. In addition to typical bacterial infections, people with CF more commonly develop other types of lung disease. Among these is allergic bronchopulmonary aspergillosis, in which the body's response to the common fungus *Aspergillus fumigatus* causes worsening of breathing problems. Another is infection with *Mycobacterium avium complex* (MAC), a group of bacteria related to tuberculosis, which can cause further lung damage and does not respond to common antibiotics.

Mucus in the paranasal sinuses is equally thick and may also cause blockage of the sinus passages, leading to infection. This may cause facial pain, fever, nasal drainage, and headaches. Individuals with CF may develop overgrowth of the nasal tissue (nasal polyps) due to inflammation from chronic sinus infections. These polyps can block the nasal passages and increase breathing difficulties.

- Produces mucus, need CFTR to water it down in the bronchial tree, need to liquefy secretions, but you don’t have the ability to do it, it doesn’t allow the lungs to move the mucus,
- Respiratory distress – clogged ducts, dried thick sticky mucus
- Leads to infections – major cause of death
- Sinuses and nasal polyps develop early in children, usually adults don’t get them until 40’s,
- HCL comes up and burns and irritates and causes GERD

b. Gastrointestinal tract (small and large intestines)

- Stools are thick, fat in the stools (steatorrhea), smelly, hard to pass cause it is big and bulky undigested food, undigested, rotted, smells terrible, no absorption of nutrients, small arms and legs, no muscles, no protein, look like ethipians, no proteins in arms and legs, bulky stools – big belly
- 2 things can happen as result
  - Newborn – not digesting anything yet, miconium usually black in color and sticky, but now stickier like glue – *muconium ilias* (obstruction of bowel caused by thick sticky plug like miconium), this is how it is usually detected, they may start vomiting and are started, restless, irritable, no nutrition, no peristalsis, bowel obstruction
  - As stools become larger, they could be constipated, it could cause *rectal prolapsed*, the bowel will push out of the rectum, very scary, lub finger and push it back in, and let peristalsis eliminate it, this is more like 2-3
years old, it could dry out if left out, you have to push it back through in internal and external sphincters
  o  Prevent it with pancreatic enzymes
• Some people may not have it so bad, they may have a little bit of NACL, and may not present until school age
c. Pancreas
The thick mucus seen in the lungs has a counterpart in thickened secretions from the pancreas, an organ responsible for providing digestive juices which help break down food. These secretions block the movement of the digestive enzymes into the duodenum and result in irreversible damage to the pancreas, often with painful inflammation (pancreatitis). The lack of digestive enzymes leads to difficulty absorbing nutrients with their subsequent excretion in the feces, a disorder known as malabsorption. Malabsorption leads to malnutrition and poor growth and development because of calorie loss. Individuals with CF also have difficulties absorbing the fat-soluble vitamins A, D, E, and K. In addition to the pancreas problems, people with cystic fibrosis experience more heartburn, intestinal blockage by intussusception, and constipation. Older individuals with CF may also develop distal intestinal obstruction syndrome when thickened feces cause intestinal blockage.

Thickened secretions also may cause liver problems in patients with CF. Bile secreted by the liver to aid in digestion may block the bile ducts, leading to liver damage. Over time, this can lead to cirrhosis, in which the liver fails to rid the blood of toxins and does not make important proteins such as those responsible for blood clotting.

• Protein plants itself, it is in exocrine glands in ducts, but instead it is in walls of the membranes in the linings of pancreas (digestive enzymes amalayze, lipase and tripsin) for carbs, fat and protein metabolism, the membrane does not allow for Nacl and water to get int there to thin out the secretion and therefore it is thick and plugged up in the lining of the ducts, the protein is not there CFTR to keep the secretions thin, instead thick mucus plugs block it, pancreatic digestive enzymes go into the common duct and the duodenum to break down food into elements that can be absorbed
• Back up of the ducts
• ADEK not absorbed into body with out pancreatic enzymes, islets of langerhans later causes diabetes – diabetes is acquired because the pancreats shuts down and doesn’t work anymore
d. Reproductive tract
Prior to prenatal and newborn screening, cystic fibrosis was often diagnosed when a newborn infant failed to pass feces (meconium). Meconium may completely block the intestines and cause serious illness. This condition, called meconium ileus, occurs in 10% of newborns with CF. In addition, protrusion of internal rectal membranes (rectal prolapse) is more common in CF because of increased fecal volume, malnutrition, and increased intra-abdominal pressure due to coughing.

• Vas def is so thickened that it can’t pass semen,
• Female – too thick to allow the sperm motility, may not make it through
  o  Delayed menarchy, may not get them until late or at all, menstrual irregularity (secretion of hormones)
Usually can’t conceive, look at hormone levels, viscosity of mucus
We need to talk to them about fertility
They can still have sex just fine, but they won’t be able to conceive

4. List the assessments and diagnostic data required to confirm cystic fibrosis (include normal values).
   a. Salty taste to the skin. People with cystic fibrosis tend to have higher than normal amounts of salt (sodium chloride) in their sweat. This may be one of the first signs parents notice because they can taste the salt when they kiss their child.
   b. Blockage in the bowels.
   c. Foul-smelling, greasy stools.
   d. Delayed growth.
   e. Thick sputum. It’s easy for parents to overlook this sign because young children tend to swallow their sputum rather than cough it up.
   f. Coughing or wheezing.
   g. Frequent chest and sinus infections with recurring pneumonia or bronchitis.
   h. Protrusion of part of the rectum through the anus (rectal prolapse). This is often caused by stools that are difficult to pass or by frequent coughing.
   i. Enlargement or rounding (clubbing) of the fingertips and toes. Although clubbing eventually occurs in most people with cystic fibrosis, it also occurs in some people born with heart disease and other types of lung problems.
   j. Growths (polyps) in the nasal passages
   k. Cirrhosis of the liver due to inflammation or obstruction of the bile ducts
   l. Displacement of one part of the intestine into another part of the intestine (intussusception) in children older than age 4
   m. Failure to grow
   n. Bulky and greasy stools (steatorrhea)
   o. Frequent respiratory infections

5. Assessments – as listed in class
   a. Mirconium ilias and vomiting
   b. Frequent resp infections
      i. Emphesema
      ii. Hear crackles because of all the mucus stuck
      iii. Hear friction rub with pneumonia
      iv. Could hear wheezing with secretions obstructing airways
      v. Could hear gurgles too
   c. Stool
      i. Undigested and smelly
      ii. Fat in stools
      iii. Prolapsed rectum in childhood
   d. No muscles
   e. Failure to thrive – grow, develop, weight gain
   f. Persistant coughing and mucus
   g. Protuberant abdomen
   h. Barrel chesting – chronic symptom usually, but CF child develops it in infancy and early toddlerhood
   i. Clubbing develops quickly
   j. Sputum
      i. Thick and yellow
ii. Bleeding – infection, will bleed into the lungs when you have infection

k. Fatigue – no energy bc no nutrition or oxygenation

l. Viracious appetite, the more they eat, the bigger their belly, but still malnourished, they can’t store any nutrients

m. Salty skin – instead of NACL going to thin things out, it goes out through the skin, mom’s taste saltiness

6. Diagnosis
   a. Sweat Chloride Tests – 2 times (best give away)
   b. Fecal analysis - Stool collection – all stools for 72 hours
   c. Pulmonary function studies on a school age child – PEF, rule out allergies and asthma
   d. ABG’s
   e. Aminocentesis
   f. DNA testing
   g. Sputum testing – will be thick, you may see organisms

7. Explain each treatment modality required for cystic fibrosis.
   a. Pulmonary hygiene
      • Chest PT 2 times a day
         o Bronchodilators. Use of medications such as albuterol, which can be delivered by an inhaler or a nebulizer, may help keep open the bronchial tubes by clearing thick secretions. At least 2 times a day
         o Mucalytics – an enzyme that breaks down the protein, Mucomist and pulmozime
         o Bronchial airway drainage. 20 minutes on each lobe of the lung, specialized percussion vests filled with air. Purse lipping and huf coughing, opens alveoli’s to create negative pressure and pushes them open.
            ▪ People with cystic fibrosis need a way to physically remove thick mucus from their lungs. This is often done by manually clapping with cupped hands on the front and back of the chest — a procedure that’s best performed with the person’s head over the edge of the bed so that gravity helps clear the secretions.
            ▪ In some cases an electric chest clapper, known as a mechanical percussor, is used. An inflatable vest that vibrates at high frequency also can help people with cystic fibrosis cough up secretions. Many adults and children with pulmonary cystic fibrosis need to have bronchial airway drainage at least twice a day for 20 to 30 minutes. Older children and adults can learn to do this themselves, especially if they use mechanical aids, such as vests and percussors. Young children need the aid of parents, grandparents or older siblings
         o Postural drainage
            ▪ Roll around, wheel barrel, cotton ball races, put them on lap and see if they can reach things
         o Flutter valve – vibrates thoracic cavity and releases mucus, you can feel the flutter on their chest, can be used in pneumonia (high frequency)
      • Physical exercise – games, roll around on floor, swim, need to be active
      • Low dose inhaled antibiotic
      • Goals – to clear secretions, prevent infection, get nutrients to grow
b. Pancreatic enzymes

- **Oral enzymes and better nutrition.** Cystic fibrosis can cause you to become malnourished because the pancreatic enzymes needed for digestion don't reach your small intestine, preventing food from being absorbed. As a result, you may need many more calories than you otherwise would. Supplemental high-calorie nutrition, special fat-soluble vitamins and enteric-coated oral pancreatic enzymes can help you maintain or even gain weight.

- **Enzymes** – 5-6 with meals, 1-2 with snacks with every meal, every time they eat, so it can break down the food, can be mixed in with food, can be put in milk of formula (don’t inhale any crushed meds, stools should then look normal and non-foul smelling
  - Pancrease
  - Creon
  - Ultrace
  - Viocase
  - Cotyzuime
  - Based on:
    - Weight Gain – they will be getting growth, they will not be hungry all the time
    - Normal stools should then look normal and non-foul smelling, they will be getting growth, they will not be hungry all the time

c. Nutritional supplements

- As a result, you may need many more calories than you otherwise would. Supplemental high-calorie nutrition, special fat-soluble vitamins and enteric-coated oral pancreatic enzymes can help you maintain or even gain weight.

- **ADEK** – fat soluble vitamins

- **Regular dietary volume**

- **More salt, replace Na and Cl electrolytes when in hot temps or exercising**

- **Low fat, high protein diet**

- **If they don’t eat – use a gastric tube, nutrients while they sleep at night so it doesn’t interfere with activities**

- **Supplemental milkshakes – with a special protein that doesn’t thicken mucus – Pulmacare, not at mealtime cause then they won’t eat, they need to eat and then have the shake in between meals**

d. Medication therapy (classification, action, etc.)

- **Pain relievers.** Ibuprofen (Advil, Motrin, others) may slow lung deterioration in some children with cystic fibrosis. (but can cause GI irritation)

- **Antibiotics.** Newer antibiotics may more effectively fight the bacteria that cause lung infections in people with cystic fibrosis. Among these are aerosolized antibiotics that send medication directly into airways. One of the major drawbacks of long-term use of antibiotics is the development of bacteria that are resistant to drug therapy. In addition, using antibiotics over a long period of time can lead to fungal infections of the mouth, throat and respiratory tract.
• **Tobramycin, Gentamycin, Tocarcylin** (strong drugs)
• **Sulfa drugs** – bactrum, to keep bacteria count down
• **GI** – go lightly prep, colight, stool softeners, steroid therapy to reduce inflammation
  o **DON’T Give** – cough suppressants, antihistamines
  o **Can Use** – decongestants, corticosteroids intra nasal, histamine receptor (prilosec, nexium)
• **Immunization** – yearly flu and pneumococcal
• **Gene therapy** – working with stem cells for future, grow new lung issue without the mutation

8. Summarize the nursing implementations indicated for clients with cystic fibrosis (include teaching).
   • **Keep your child's immunizations up to date.** In addition to other usual childhood vaccines, this includes the pneumococcal and influenza vaccines. Cystic fibrosis doesn't affect the immune system, but children with cystic fibrosis are more likely to develop complications when they become sick.
   • **Encourage your child to lead as normal and active a life as possible.** Exercise is extremely important for people of all ages who have cystic fibrosis. Regular exercise helps loosen mucus in your airways and strengthens your heart and lungs. And for many people with cystic fibrosis, participating in sports can improve confidence and self-esteem. It isn’t necessary to take part in an organized sport or take classes at a gym. Anything that gets you moving, including walking and biking, can help.
   • **Make sure your child eats a healthy diet.** Be sure to discuss your child's dietary needs with your doctor or a nutritionist.
   • **Use nutrition supplements.** Provide the fat-soluble vitamin supplements and pancreatic enzymes your child needs to stay as healthy as possible.
   • **Emphasize liquids.** Encourage your child to drink plenty of liquids to help loosen the mucus. This is especially important in the summer when children are active and tend to lose a lot of fluids.
   • **Eliminate smoke.** Don't smoke in your home or car, and don't allow other people to smoke around your child. Secondhand smoke is harmful for everyone, but especially for people with cystic fibrosis.
   • **Encourage hand washing.** Teach everyone in your family to wash their hands thoroughly before eating, after using the bathroom, when coming home from work or school, and after being around a person who is sick. Hand washing is the best way to protect against infection.
   • **Teaching, supervision, referrals**
   • **IV access** – venous access device so it is there when they need it
   • **O2 equipment** – they need to know how to clean it
   • **Standard precautions** – even when you are in your own home, it can be lethal to a CF person
   • **CF Foundation** – they can get you equipment and finances
   • **Siblings** – will feel jealous and won't get as much attention, may feel neglect, they could feel the disease, anger at parents and other sibling, reject the other sibling, negative behaviors to get in trouble and get parents attention, there needs to be time for the normal family, animocity and they may even think they wish he’d die, mom and dad would have more time for them, guilt
• **Huband and Wife** – no time for each other, no vacation time, up with the child at night, family crisis on daily basis, someone is unhappy, increase in divorces, 1 parent takes off

• **Psychologist**

• **Pschiatrist**

9. Describe the complications of cystic fibrosis

• **Respiratory complications**

Frequent complications of cystic fibrosis are chronic respiratory infections, including pneumonia, bronchitis, chronic sinusitis and bronchiectasis — an abnormal dilation of the walls of the bronchial tubes that makes it more difficult to clear your airways. Asthma can result from chronic inflammation of the bronchial lining.

- **Respiratory infections** are common because thick mucus blocks the airways and provides an ideal breeding ground for bacteria. The most common infective agent in people with cystic fibrosis is *Pseudomonas aeruginosa* — a bacterium that can cause increased inflammation of the respiratory tract. Although antibiotics can decrease the frequency and severity of attacks, the bacteria are never completely eradicated from the airways and the lungs. On the other hand, *P. aeruginosa* rarely causes pulmonary infections in healthy people and isn’t considered contagious.

- **People with cystic fibrosis** may also develop bleeding from the lungs causing them to cough up blood (hemoptysis), respiratory failure or collapsed lung (pneumothorax) — a condition in which lung air leaks into the chest cavity through a small hole that forms in the lung’s outer layer. Lung disease eventually may cause the lower right chamber (right ventricle) of the heart to fail. Ultimately, complications from lung problems prove fatal for many people with cystic fibrosis.

  - **Bleeding** - +200 ml blood, cauterize vessel
  - **Pneumothorax** – collapsed lung, pulmonary blebs
  - **Cor Pulmonali** – right sided heart failure due to pulmonary congestion
  - **Diabetes**
  - **Chronic sinusitis**
  - **Nasal polyps**
  - **Cirrhosis of the liver** –
  - **Portal hypertension** –
  - **Intestinal obstruction** –
  - **Bronchitis/pneumonia** –
  - **Lung transplant** – rejection is a problem, prolongs life for 5-7 years, still have pancreatic problems

• **Nutritional complications**

In addition, cystic fibrosis makes you prone to chronic diarrhea and severe nutritional deficiencies. That's because thick secretions obstruct the ducts in your pancreas, preventing enzymes that digest fats and proteins from reaching your intestines. These secretions also prevent your body from absorbing the fat-soluble vitamins A, D, E and K.

- **Cystic fibrosis** affects the pancreas and because the pancreas controls the level of sugar in your blood, up to one in five people with cystic fibrosis may develop cystic fibrosis-related diabetes. In addition, the bile duct, the duct that carries bile from
your liver and gallbladder to your small intestine, may become blocked and inflamed, leading to liver problems, such as cirrhosis.

- **Reproductive complications**
  Cystic fibrosis also affects the reproductive system. Because thick secretions often block the tube connecting the testes and prostate gland (vas deferens), many men with cystic fibrosis are infertile. But certain fertility methods and surgical procedures may sometimes make it possible for men with cystic fibrosis to become fathers.
  - Although women with cystic fibrosis may be less fertile than other women, it’s possible for them to conceive and to have successful pregnancies. Still, pregnancy can worsen the signs and symptoms of cystic fibrosis, so be sure to discuss the possible risks with your doctor. Using oral contraceptives also can sometimes aggravate certain symptoms of cystic fibrosis. Talk to your doctor about your birth control options.