Cystic Fibrosis

An Honors Thesis (ID 499)

By

Lori Mattix

Thesis Director

Mrs. Carolyn S. Caldwell

Carolyn Sue Caldwell

Ball State University

Muncie, Indiana

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OBJECTIVES FOR INDEPENDENT STUDY ON CYSTIC FIBROSIS

I will work with children who have cystic fibrosis.
   a) hospital setting
   b) children in the community
   c) find out their feelings on it—how it affects them/
      their friends/their family

I will understand how the family is affected and some of their
   a) parents
   b) grandparents
   c) siblings

I will know the economic effects of cystic fibrosis.
   a) hospital
   b) physician
   c) drugs
   d) treatments
   e) equipment

I will research any new technological findings related to CF.*
   a) cure
   b) cause
   c) new treatments

I will know where these families can go to get help (financially,
   socially) and how hard it is to get this help.
   a) government agencies
   b) community agencies
   c) private agencies
   d) Cystic Fibrosis Foundation

I will understand the etiology and incidence of CF.

I will be able to complete a general nursing care plan for a
   patient with cystic fibrosis.

I will know about the diagnostic procedures for cystic fibrosis.
   a) types
   b) cost
   c) accuracy

*Cystic Fibrosis
INTRODUCTION

When I came to Ball State in 1974, I decided to join a sorority. Alpha Chi Omega was the one I chose. Alpha Chi’s national philanthropic is cystic fibrosis (CF). Although I collected money for it, I really had no idea what CF was.

When I started nursing, I learned about cystic fibrosis. I was very disappointed that so few people seemed to know about this disease. For this reason, I decided to do my honor’s thesis on cystic fibrosis.

Someday, I’d like to be a nurse practitioner in a CF center.

Cystic fibrosis is a generalized hereditary disorder of children, adolescents, and young adults in which there is a widespread dysfunction of the exocrine glands. In fully manifested cases, chronic pulmonary disease, pancreatic enzyme deficiency and abnormally high sweat electrolytes are present. The disorder is characterized by differing degrees of involvement of affected organs and glandular systems, resulting in variation in the clinical expression of the disease.2

1 Hereafter, CF will be used to refer to cystic fibrosis.

The incidence of cystic fibrosis is believed to be between one in one thousand and one in sixteen hundred live births, with the carrier incidence between three and six percent of the population. This makes cystic fibrosis one of the more common of the serious chronic diseases of childhood and adolescence. CF is the most frequent lethal genetic disease among white children. Today fifty percent of cystic fibrosis children live past eighteen years of age.\(^3\)

With the realization that cystic fibrosis children can survive to adulthood if diagnosed early and given proper treatment, the Cystic Fibrosis Foundation is making a strong push to have the disease become better known.

\(^3\)Ibid., p. 2.
Figure 1  GENETIC POSSIBILITIES FOR EACH PREGNANCY
WHEN BOTH PARENTS ARE CARRIERS

Sperm I and Egg III both carry the CF gene.

Father  

\[ \text{Sperm I} \quad \text{Sperm II} \]

Mother  

\[ \text{Egg III} \quad \text{Egg IV} \]

Child A  

Child B  

Child C  

Child D

Child A: Sperm I fertilizes egg III. The child receives two genes for CF and therefore inherits CF.

Child B: Sperm I fertilizes egg IV. The child inherits one gene for CF and, thus, is a carrier, like the parents.

Child C: Sperm II fertilizes egg III. The child receives one gene for CF and also is a carrier.

Child D: Sperm II fertilizes egg IV. The child inherits no genes for CF. He or she therefore does not inherit CF, is not a carrier and could not transmit the CF gene to future generations.

4 The Genetics of Cystic Fibrosis, Cystic Fibrosis Foundation, Atlanta.
PATHOPHYSIOLOGY

Cystic fibrosis is sometimes referred to as the great impersonator. It is a great simulator of other diseases because the symptoms are diverse and not always clear. There is widespread dysfunction of the exocrine glands. Exocrine glands are glands which secrete their products through ducts. Due to the dysfunction, excessive amounts of thick mucus are produced. This thick mucus can lead to the damage of some organs. In 1936, Dr. Guido Fanconi first reported cystic fibrosis of the pancreas. Two years later, in 1938, Dr. Dorothy Anderson described CF as a separate disease. It was not until 1944, however, that cystic fibrosis was found to be a disease of the exocrine glands rather than of the pancreas. 5

The amount of involvement of various organs varies with each individual, however, the lungs seem to be one of the more seriously affected areas. The production of thick, tenacious bronchial mucus causes the problem in CF lungs. This mucus cannot be easily propelled by ciliary action. The accumulation of these secretions causes narrowing of the airways. As the cystic fibrosis progresses, the areas of narrowed airways increases. Obstruction of the airways causes a resis-

tance to airflow, particularly in expiration, since the lumen of bronchi are narrower during expiration than inspiration. This obstruction, in turn, leads to obstructive emphysema. When the obstruction becomes widespread, alveolar ventilation is impaired.6

Figure 2 Obstructive Lung Disorder

As the alveoli hyperinflates, there is an increase in the anteroposterior diameter, as well as a low, flat diaphragm.8

Due to the obstruction of airways, stagnant mucus collects in the lungs. This provides a good growth medium for bacteria. Staphylococcus aureus is the most common invader. After much antibiotic treatment, Pseudomonas aeruginosa and Klebsiella infect the lung. These bacteria are not sensitive to most antibiotics. A purulent bronchitis is then superimposed on the obstructive emphysema further impairing alveolar ventilation.9


7 Ibid.


The pulmonary disorder associated with cystic fibrosis affects the cardiovascular system. Due to the damage of the alveolar capillaries in the lungs, the right ventricle must pump harder in order to get blood into the lung. The need for increased pumping power causes the right ventricle to hypertrophy. This leads to right sided heart failure or cor pulmonale. Too small of a blood supply for too large of a ventricle leads to failure. 10

The second major organ system affected is the digestive system. Patients with cystic fibrosis cannot digest fat. This is due to the absence of pancreatic lipase, the enzyme which breaks down fat. Lipase is absent because the duct which carries the enzyme is filled with the thick mucus. "There is a 50-75 percent absorption rate of fats. (Normal is 95-97 percent) Absorption of any fat in the absence of pancreatic lipase has been explained by the production of small amounts of intestinal lipase and/or absorption through emulsification." 11

Since much of the fat is not absorbed, it passes through the intestine and results in steatorrhea. Steatorrhea is bulky, foul smelling stools. In addition to this, the CF patients also have azotorrhea and excrete increased


amounts of nitrogen in their stool as a consequence of the absence of trypsin. While normal subjects absorb between 97-98 percent of dietary protein, cystic fibrosis patients may absorb only 40-80 percent.\footnote{Henry L. Barnett, M.D. and Arnold H. Einhorn, M.D., Pediatrics, 15th ed. (New York: Appleton-Century-Crofts, 1972), p. 417.}

Since the patients do not absorb enough fat, the absorption of vitamins A, D, and E is impaired. Therefore, it is important to give vitamin supplements.

The normal sweat gland produces an isotonic secretion which is modified by reabsorption of both water and electrolytes. This is also true in cystic fibrosis. The defect appears to be in the duct where increased reabsorption of sodium and chloride occurs. Water reabsorption in the excretory duct is not increased. Thus the sodium and chloride concentrations in the sweat of cystic fibrosis patients are two to five times the normal values.\footnote{Thomas F. Boat, M.D., Bernard Boxerbaum, M.D. Carl F. Doershuk, M.D., LeRoy W. Matthews, M.D., J. Primiano, and Robert C. Stern, M.D., "Cystic Fibrosis - An Obstructive Pulmonary Disease of Children and Adults," Department of Pediatrics Case Western Reserve University School of Medicine, (Cleveland, 1976), p. 3.}

There are several possible complications which can occur in a patient with cystic fibrosis.

Meconium ileus is the earliest manifestation of cystic fibrosis. It occurs in ten to fifteen percent of newborns with CF.\footnote{Ibid. It occurs when the bowels are obstructed by putty-like intestinal secretions. The results are abdominal
distension, vomiting, and inability to defecate. Enemas with pancreatic enzymes may be tried, but usually surgery is necessary at two to three days of age to relieve the obstruction. The fact that the infant has this does not indicate that the disease will be severe.15

Rectal prolapse, which occurs in about twenty percent of inadequately treated cases of cystic fibrosis, is usually recurrent and probably due to both poor perineal muscle tone and the passage of bulky, oily stools.16

Diabetes mellitus has been reported as a complication. This could possibly be due to the fibrosing of the pancreas thus leading to the destruction of some Islets of Langerhaans. Although the incidence of severe diabetes is low, the glucose tolerance test is impaired in many cases and the amount of insulin produced during the test is lower than normal.17

Heat prostration due to excessive loss of sodium chloride in the sweat may occur any time, but is more commonly

seen in the summer. The diagnosis is confirmed by lab tests showing severe hyponatremia. Signs and symptoms are circulatory collapse with hyperpyrexia, convulsions, and eventually death if not treated.\textsuperscript{18}

Nasal polyps are common and require surgical removal when they cause obstruction or chronic nasal infection. Although the etiology is unclear, allergies and infections are believed to play important roles. Nasal polyps tend to be recurrent.\textsuperscript{19}

As more patients survive to older ages, a greater number of CF patients develop cirrhosis of the liver. At first the liver enlarges, but eventually it shrinks in size. The cirrhosis results from mucus deposits in the liver causing fibrosis. Development of portal hypertension in approximately five percent of cystic fibrosis patients is manifested by: esophageal varices with resultant hematemesis or melena, progressive enlargement of the spleen, and thrombocytopenia. Symptoms of hepatic failure are uncommon. Severe chronic or acute symptoms may require surgical intervention.\textsuperscript{20}

Most males with cystic fibrosis are infertile. There are no vas deferens. Instead the spermatic cord consists of

\textsuperscript{18} Ibid.

\textsuperscript{19} Carl F. Doershuk, M.D., Guide to Diagnosis and Management of Cystic Fibrosis, (Atlanta, GA: National Cystic Fibrosis Research Foundation, 1971), p. 11

fibrous strands and smooth muscle. Also the tail of the epididymis is usually atrophic. The spermatic ducts end blindly.\textsuperscript{21}

There are several pulmonary complications which can occur in the patient with cystic fibrosis:

- atelectasis - occurs when mucus plug prevents aeration of the lungs
- pneumothorax - occurs in advanced disease and is felt to occur due to rupture of surface islet of pulla
- hemoptysis - associated with severe infection\textsuperscript{22}

Although cystic fibrosis has symptoms similar to other diseases, certain signs are indicative of CF. A summary of these symptoms follows.


Primarily Due To Pulmonary Lesion

Symptoms and Signs of Low-Grade Chronic Pulmonary Infection

**Early**
- Dry, hacky, non-productive cough
- Increased respiratory rate
- Prolonged expiratory phase of respiration
- Decreased activity

**Moderate**
- Increased cough with sputum production
- Rales, musical rhonchi, scattered or localized wheezes
- Repeated episodes of respiratory infection
- Signs of obstructive lung disease
  - Increases anteroposterior diameter
  - Diminished area of cardiac dullness
  - Depressed diaphragm
  - Palpable liver border
- Decreased appetite—may still be good but not voracious
- Failure to gain or grow, or weight loss
- Decreased exercise tolerance

**Advanced**
- Chronic, paroxysmal, productive cough, often associated with vomiting
- Increased respiratory rate, shortness of breath on exertion, orthopnea, dyspnea
- Diffuse and localized rales and rhonchi
- Signs of marked obstructive lung disease
  - Marked increase in anteroposterior diameter—barrel chest, pigeon breast
  - Limited respiratory excursion of thoracic cage
  - Depressed diaphragms
  - Hyper-resonance over entire chest
  - Decreased exchange
- Noisy respiration—wheezing, bubbling, audible rales
- Marked decrease in appetite associated with weight loss
- Growth failure—stunting
- Muscular weakness—flabby
- Cyanosis
- Digital clubbing
- Rounded shoulders, forward position of head, poor posture
- Fever, tachycardia, toxicity
- Hemoptysis

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continued

Atelectasis
Pneumothorax
Lung abscess
Signs of cardiac failure—edema, enlarged tender
liver, venous distention
Visual impairment and eye ground changes

Primarily Gastrointestinal

Pancreatic and Nutritional
Meconium ileus
Intestinal obstruction
Fecal masses
Poor weight gain despite voracious appetite
Vitamin deficiencies
Distended abdomen
Three to four bulky, greasy, floating foul-smelling stools per day
Rectal prolapse
Palpable loops of bowel, especially in right lower quadrant
Cramps and excessive foul flatus
Malnutrition, poor muscle tone, small flabby muscles, lack of subcutaneous fat
Hypoproteinemia with generalized edema

Hyponatremia and Hypochloremia
Loss of sodium and chloride by sweating
Mild—forehead tastes salty
Severe—muscle cramps, weakness, shock

Biliary Cirrhosis and Portal Hypertension
Firm, nodular liver—often palpable in midline
Splenomegaly
Hypersplenism—decreased white blood cells and platelets, anemia
Hematemesis and melena from esophageal varices

Genital Tract Involvement

Aspermia, absence of vas deferens
Cervical polyps, increased viscosity of mucus

24 Ibid., p. 22.
Chapter II

DIAGNOSTIC PROCEDURES/LABORATORY STUDIES
The most reliable diagnostic test for cystic fibrosis is the quantitative analysis of sweat for chloride and sodium. No other disease that can be confused with CF on a clinical basis has a constant elevation of sweat electrolyte levels. Children with CF will have a positive sweat test from the day of birth. This test is over 99 percent reliable. The diagnostic sweat test consists of four steps:

1. Stimulation of local sweat glands by the pilocarpine iontophoresis method. An electric current (2.5-3.0 MA) drives the pilocarpine into the skin for five to ten minutes to induce local sweating. Some people inject the drug intradermally instead.

2. Collection of the specimen. The forearm is the preferred area for collection. The site should be clean. The specimen may be absorbed onto gauze or filterpaper previously weighed so that the amount of sweat can be accurately determined. The usual time for sweat collection is thirty minutes. Care is needed to avoid contamination, evaporation, and errors in weighing the sample.

3. Quantitative analysis. The chloride concentration is determined by a titrimetric and the sodium concentration is determined on the flame photometer. These values should be within ten to fifteen percent of each other; wider discrepancies should arouse suspicion concerning the analysis or the calculations of the results. Fifty milligrams of sweat is the minimum amount of sweat which should be tested.

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4. Interpretation of sweat test. The finding of a sweat chloride above concentration 60 milliequivalents/liter or a sodium above 70 milliequivalents/liter is consistent with the diagnosis of cystic fibrosis. If the results show the patient to be borderline (sodium and/or chloride between 50-70 milliequivalents/liter) the sweat test should be repeated and other laboratory tests should be carried out.

One point which must be emphasized is that the sodium and chloride levels do not affect the severity of the disease.

Pancreatic function tests used to be the main diagnostic test for cystic fibrosis. With the advent of the sweat test, however, this test has lost its popularity. In order to perform this test, the duodenum is intubated. The pancreas is then stimulated by giving intravenous secretion and pancreozymin. At timed intervals the duodenal secretions are aspirated. Testing is done to see if trypsin is found in the duodenum. If trypsin is not found, it means that there is a blockage in duct between the pancreas and the duodenum. This is indicative of cystic fibrosis.

An indirect test of pancreatic function is a stool test. If trypsin is absent in the intestine, it will not be present in the stool.

Pulmonary testing may be used diagnostically, but usually it is used for follow-up assessments. Chest x-rays show obstructive phenomena with irregular aeration and patchy atelectasis. Overinflation is also commonly seen.

26 Ibid., pp. 27-31.
As the disease progresses, the anteroposterior diameter increases, the diaphragm is depressed and areas of infiltration are seen.

Early diagnosis is the key. The sooner treatment is started on cystic fibrosis patients, the longer their life expectancy.

Figure 4

CHART 27
Diagnosis of Cystic Fibrosis

<table>
<thead>
<tr>
<th>System</th>
<th>Sweat Gland Involvement</th>
<th>Pulmonary Involvement</th>
<th>Pancreatic Involvement</th>
<th>Genetic Involvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinical</td>
<td>salt depletion in hot weather</td>
<td>cough obstruction</td>
<td>streatorrhea malnutrition</td>
<td>family history</td>
</tr>
<tr>
<td>Test</td>
<td>sweat test</td>
<td>chest x-ray</td>
<td>duodenal fluid or stool assay</td>
<td>sweat test</td>
</tr>
<tr>
<td>Observed</td>
<td>increased sweat electrolytes</td>
<td>generalized overinflation infection</td>
<td>absence of pancreatic enzymes</td>
<td>siblings or first cousins with CF</td>
</tr>
</tbody>
</table>

27 Ibid., p. 42.
As the cystic fibrosis progresses other lab tests may be performed to check for complications.

A vectorcardiogram demonstrates right ventricular hypertrophy due to cor pulmonale.

Sputum analysis is done to check for effectiveness of antimicrobial treatment. Also to prescribe antibiotics, this test may be necessary.

Pulmonary function tests are used to assess the disease process. The obstructive disease is shown through overinflation, as demonstrated by an increase in the residual volume, an increase in the total lung capacity, and a decrease in vital capacity. Expiratory flow rates are decreased. Blood gases vary from normal to the extremes of hypoxemia and finally hypercapnia.²⁸

The above lab tests are often done regularly at Cystic Fibrosis Centers in order to compile data for statistics and research.

Parent teaching plays an important part in cystic fibrosis detection. The Cystic Fibrosis Foundation has movies such as "Kiss Your Baby" which show the parents what type of things to look for to see if their child has CF. Some examples are salty taste when kissed, coughing followed by vomiting, coughing and wheezing, failure to gain weight with a good appetite, wasting buttocks, pot belly, frequent

bulky, foul-smelling stools and recurrent pulmonary infections. By coordinating the efforts of parents and physicians in detecting cystic fibrosis in the infant, the prognosis is much better.

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Chapter III

TREATMENT
TREATMENT

Physical Therapy for Bronchial Obstruction

Cystic fibrosis children have excess mucus in their lungs. Unless this mucus is cleared out, infection can result.

Nembulization therapy is very important in the treatment of pulmonary problems. It consists of sending small particles of moisture into the air. The tasks which nembulization performs are:

1. to humidify the respiratory tract
2. to wet and thin secretions in the respiratory tract
3. to deposit medications on the mucosal surface

Propylene glycol is the substance used in most nembulizers. It helps to stabilize particle size and prevent complete evaporation. Clean equipment for nembulization therapy is necessary. If the equipment is contaminated, there is a chance of conveying an infection to the CF child. 30

Inhalation therapy involving the nembulization and direct inhalation of medications is called intermittent aeresol therapy. The purpose of this aeresol therapy is to deposit substances in the the tracheobronchial tree. Here are the various types of therapy used:

hand firmly on the patient's chest wall. Vibration is performed during exhalation, with the patient exhaling as slowly and completely as possible. Five exhalations and vibrations per position is sufficient.  

Frequency, duration and positions of percussion and postural drainage should be determined individually. Auscultation, x-ray, and the individual's ability to assume position can determine this.

When percussion and postural drainage are used prophylactically, the lower lobes are usually drained first, followed by the middle lobe and finally the upper lobes. This order is reversed in small infants, post-operative patients, or patients whose predominant position is recumbent.  

See Appendix A.

Mist tent therapy is used by patients with significant pulmonary involvement. It is also felt to be beneficial in prophylactic therapy. Mist tent therapy is used to humidify inspired air and to deposit particulate water in the respiratory tract.

Physical activity is a very good method of keeping the lungs free from accumulation of secretions. Running, swimming and active play loosen the secretions and encourage coughing.

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32 Ibid., p. 54-55.
33 Ibid., p. 53.
34 Ibid., p. 49.
Drug Therapy

Antibiotics

A. Penicillins and Synthetic Penicillins

Action: Inhibit synthesis of bacterial cell walls, probably by interfering with the biosynthesis of mucopeptides and preventing linkage of structural components of the cell wall. Most penicillins are much more active against gram-positive than gram-negative bacteria.

Indications: staphylococcal infections; hemophilus influenza; group A streptococci

Adverse effects: skin rash; nausea; vomiting; central nervous system toxicity; excitation and convulsions if large dose given; anaphylactic shock/allergic reactions; diarrhea; black 'hairy' tongue

Nursing Implications: Watch for signs of superinfection. With some of these drugs, a false positive reaction with Clinitest may occur, but it will not occur with Tes-Tape. Ask patient if any previous allergic reaction to any penicillins. Do not give orally with food.

Dosages: See Appendix B

B. Cephalosporins

Action: Inhibit synthesis of bacterial cell walls, probably by interfering with the biosynthesis of mucopeptides and preventing linkage of structural components of the cell wall. Most penicillins are much more active against gram-positive than gram-negative bacteria.

Indications: staphylococcal infections; hemophilus influenza

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37 Ibid.
Indications: both gram negative organisms and staphylococcus aureus

Adverse reactions: Anorexia, nausea, vomiting, diarrhea, glossitis, local irritation after IM injection, rashes; anaphylactic reactions, phototoxicity; liver damage if large doses

Nursing Implications: Administration during pregnancy or in children under seven years could cause discoloration of teeth and depression of bone growth. Watch for signs of superinfection. Don't give with milk products or antacids or iron products as they will form nonabsorbable compounds. May be given with meals. Protect drug from sunlight and extreme heat or humidity as they will lose potency. Tetracyclines delay blood coagulation.

Dosages: See Appendix B

E. Aminoglycosides

Action: Act upon the bacterial ribosome to induce specific misreading of the genetic code.

Indications: pseudomonas infections, gram negative organisms

Adverse effects: Ototoxic - damage both the vestibular and the auditory branches of the eighth cranial nerve. Nephrotoxic, neuromuscular blockade leading to respiratory arrest has been observed. Pain at injection site, skin rash, headache, drug fever

Nursing Implications: Observe for acoustic nerve damage tinnitus, vertigo, hearing loss, ataxia. Observe for signs of nephrotoxicity - decreased urine output, run creatinine clearance test. Decrease dosage in aged, young or debilitated patients.

Dosage: See Appendix B

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41 Ibid.

F. Macrolides

Action: Bacteriostatic by means of inhibiting protein synthesis.

Indications: staphylococcal infections

Adverse effects: nausea, vomiting, diarrhea, allergic reaction

Nursing implications: Watch for signs of superinfection. If jaundice occurs stop drug - could be due to hepatotoxicity.

Dosage: See Appendix B

G. Chloramphenicol

Action: Potent inhibitor of protein synthesis.

Indications: gram negative organisms and staphylococcus

Adverse effects: bone marrow depression, anemia, leukopenia, aplastic anemia, gastrointestinal disturbances and optic neuritis

Nursing Implications: Watch for signs of superinfections. Run periodic blood counts. Do not give to newborn as grey baby syndrome may occur - vasomotor collapse, hypothermia, rapid and irregular respirations, diarrhea, ashen color and death.

Dosage: See Appendix B

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44 Ibid.

Mucolytic Agents

Action: Reduce thickness and stickiness of purulent and nonpurulent pulmonary secretions. They are usually administered by aerosol inhalation to facilitate removal of secretions. Some mucolytic agents work only through humidification while others contain enzymes which break down the mucus.

Adverse effects: stomatitis, nausea, rhinorrhea, bronchospasm, allergy

Nursing implications: Wash patient after treatment as he may be sticky. Have bronchodilator present in case of bronchospasm.

Dosages of various types:
- Acetylcysteine (Mucomyst) 1-10 milligrams of 20 percent solution three to four times daily
- Desoxyribonuclease (Dornovac) 50,000-100,000 units three times per day

Enzymes

Action: Pancreatic enzymes reduce the wastage of dietary fat and protein as well as improved utilization of fat soluble vitamins. They come from pork pancreas usually. These enzymes take the place of the enzymes which do not reach the intestine in cystic fibrosis children.

Adverse Effects: The only problem usually encountered is allergy. It may make the stools softer and more frequent. Intestinal discomfort occurs occasionally. Too much of the enzyme may result in constipation.

Nursing implications: Assess for proper amount of enzyme replacement. Inadequate pancreatic replacement and excessive fat intake produce abdominal cramps and distension, light colored, mushy and floating stools, foul smelling flatus, oozing from rectum, rectal prolapse, or failure to thrive despite good appetite. Reassure parents that with start of enzyme therapy it is normal for the child's appetite to decrease.

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46 Ibid., pp. 394-95.
Dosages of types enzymes:

- **Cotazym capsules** (350 milligrams) 1-5 with each meal or snack
- **packets** 4-5 packets with each meal or snack
- **Viokase (beef pancreas) powder** 1.5 grams/teaspoon 4-1 teaspoon with each meal or snack
- **tablets** (325 milligrams) 4-8 tablets with each meal or snack

**Diet Therapy**

Children with cystic fibrosis have much trouble with their digestive processes due to the lack of pancreatic lipase. For this reason pancreatic enzymes are given. The amount to be given should depend upon the response of the patient to the medication.

An adequate amount of calories for growth and development must be given. Small infants may require up to 200 calories/kilogram/day. A low fat diet under one year of age should be avoided because the essential fatty acids should not be excluded during this period of rapid growth. The older child is maintained on a relatively high protein diet. The total fat in the diet is restricted, however, rigid dietary control is not necessary. The diet should be adjusted to the individual's tolerance.

---

Vitamin supplementation is an essential part of the treatment for cystic fibrosis patients. Fat soluble vitamins, especially vitamins A and D, are prescribed in twice the usually recommended dosages. Vitamin K supplement is given twice weekly for infants during the first year of life, and at any age if the patient has cirrhosis, portal hypertension, or hemoptysis. Vitamin E may be given but at this time there have not been any definite signs of vitamin E deficiency conclusively demonstrated. 

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Figure 5

DIETARY THERAPY 50

The diet should provide increased calories and protein with a variable reduction in fat as needed to satisfy the patient's appetite and requirements for growth and development. The basic four food groups should be included for a well-balanced diet.

Diet should be individualized. Selection of foods is governed by:

-the degree of pancreatic enzyme deficiency present.
-the degree of individual development (e.g. ability to chew).
-the dietary habits of the family, including economic, social, cultural and psychological factors.
-individual differences in appetite and preferences.
-modifying conditions (food allergy, diabetes, ulcer).

Calories: A 50-100 percent increase in calories above normal requirements for age is recommended.

Infants--150-200 calories per kilogram per day
Children 1-9 years--130-180 calories per kilogram per day
Males 9-18 years--100-130 calories per kilogram per day
Females 9-18 years--80-110 calories per kilogram per day

Proteins: An intake of 2-2½ times over normal is recommended.

Infants--4 grams per kilogram per day
Older children-- 3 grams per kilogram per day
Young adults--2½-3 grams per kilogram per day

Fats: Intake varies with age, total caloric intake and degree of fat intolerance. Rigid fat restriction should be avoided.

Infants
Normal: 30-60 grams per day
Moderate fat: 30-50 grams per day
Low fat: 30-40 grams per day

Older Children
Normal: 50-120 grams per day
Moderate low fat: 50-70 grams per day
Low fat: 30-50 grams per day

51 Ibid.
Chapter IV

ECONOMIC ASPECTS
ECONOMIC ASPECTS

Cystic fibrosis is a lifelong disease. The expert care and the expensive treatments make it one of the costliest diseases. Dr. Arnold Dunn, a pediatrician who adopted a CF baby, estimated his costs. A seven month hospital stay for the infant cost $36,355. An estimated $600 worth of equipment is needed for the home. The medication bill is approximately $3,000 for one year. Dr. Dunn stated that it would cost between $4,000 and $12,000 per year to keep the baby alive. These estimates are from the Washington D. C. area. 52

The first costs the family usually encounters are either hospitalization or several visits to the physician's office with complaints such as failure to thrive or bronchitis. A visit to the physician can run from $13-$16. A day in the hospital is approximately $86. These two prices do not include any prescribed medications.

Diagnostic tests make up the second major expense. So common laboratory tests and their prices include:

- duodenal fluid $7; stool test $12; chest x-ray $23.50; sweat test $12; and nose or throat culture $13. 53

52 Anita Brett, "The Doctor Who Adopted His Patient," Today's Health, March, 1974, pp. 54-61
53 Ms. , interview with Ball State Medical Technology Student at Ball Memorial Hospital, Muncie, Indiana, April 22, 1977.
These pieces of equipment are not only expensive financially, but they are also expensive in time. Each piece of equipment should be cleaned daily so that bacterial growth is kept to a minimum.

Regular visits to the physician or to the Cystic Fibrosis Center are essential to assess the health status of the CF patient. Most Centers are run on a sliding scale. The family pays only for what they can afford.\(^{55}\) Transportation to the Center or the physician's office is also an expense to the family.

Probably the most expensive aspect of CF is the drug therapy regimen. Cystic fibrotic children take around 40 pills daily. Pancreatic enzymes cost $40 for 250 packets.\(^{56}\) Depending on the meal, four to eight packets may be taken with each meal and snack. Erythromycin (250 milligrams) costs $12 for one hundred pills. At the dosage of three daily this would last one month. Organidin, a mucolytic agent is $8.50 for one pint. The usual dosage is one teaspoon four times

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\(^{54}\) Mr.\_\_\_, interview with 26 year old man with cystic fibrosis, Muncie, Indiana, September 27, 1977.

\(^{55}\) Mr.\_\_\_, interview with the Director of Indiana's Cystic Fibrosis Foundation, Indianapolis, Indiana, October 18, 1977.

\(^{56}\) Mr.\_\_\_, interview with the father of CF child, Muncie, Indiana, October 17, 1977.
daily. At this rate, the Organidin would last slightly over one week. Vitamins are an essential part of the drug therapy. The prices are as follows:

Vitamins A and D $2.29 per fifty capsules  
Vitamin E $5.28 per fifty capsules

A man with one CF daughter spent $688 on drugs for one year. This does not include visits to the physician to get the prescriptions.

A primary support service to patients with cystic fibrosis is Crippled Children's Services. This is a federal-state program providing financial aid for patient care. The type and extent of aid varies, according to the state. Presently there are only seven states which have similar programs for CF individuals over the age of 21. The Cystic Fibrosis Foundation is currently working to change this.

Government funding also helps support the Cystic Fibrosis Centers. As of June, 1976, Crippled Children's Services paid $4.2 million to families with CF children. The Pediatric Pulmonary Centers received $2.2 million to support the health care programs of CF children.

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57 Mr. , interview with an Osco pharmacist, Muncie, Indiana, November 1, 1977.

58 Mr. , interview with the father of CF child, Muncie, Indiana, October 17, 1977.


60 Ibid.

Welfare and Aid to Dependent Children (ADC) are two other places where financial aid can be received. In order to apply for any of these assistance programs, the family must go to the Child Welfare Department. Here they fill out numerous forms. Once approved (this usually takes one month), the family receives financial help which is retroactive to the date they filed for aid. The amount of aid received depends on the income of the family and the expenses.

The Cystic Fibrosis Foundation of Indiana does not give direct aid to a family but can be very helpful in referrals to places where assistance can be obtained. Often the Cystic Fibrosis Foundation can refer the family to organizations which help with the purchase of equipment. An example is that the Odd Fellows of Muncie donated a postural drainage table to a CF child. 62

Some cities offer special programs to help alleviate the cost of cystic fibrosis. The Chicago CF program allows CF families to purchase their medications at a 25–40 percent discount. 63 Indiana's Cystic Fibrosis Foundation occasionally purchases large quantities of drugs and then sells them to the CF Centers in Indiana. 64

62 Mr. , interview with a father of a CF child, Muncie, Indiana, October 17, 1977.
64 Mr. , interview with the Director of Indiana's Cystic Fibrosis Foundation, Indianapolis, Indiana, October 18, 1977.
Although there is aid to cystic fibrosis families, many people are unaware of how it can be obtained. The Cystic Fibrosis Foundation is working to solve this problem.
Chapter V

RESEARCH
RESEARCH

Backing from the federal government and from the National Cystic Fibrosis Foundation allows much research to be accomplished.

Researchers at the National Institute of Arthritis and Metabolic Diseases have done much work with cystic fibrosis. They have obtained evidence indicating that excess calcium in the salivary glands of the CF patients combines with a group of proteins to form an insoluble compound. They feel that there is a possibility that this compound is responsible for the widespread obstruction of various organs.65

One of the older theories states that the mucus produced by affected cells in a patient with CF is either abnormal or produced in a large amount. A more recent theory, however, states that the problems are caused by difficulties in the process of salt and water movement across affected cell membranes. An abnormality in the concentrations of salt and water could account for the abnormalities encountered in the mucus in cystic fibrosis, since the physical state of mucus depends on the amount of salt and water present.

Conversely, abnormal mucus could interfere with the normal movement of salts and water across the cell membrane and thus alter the fluid secreted by the gland.66

It has been found that antibodies can have a marked effect on the ability of molecules to pass through cell membranes. There is some indication that some antibodies may be lacking or present in another form in patients with cystic fibrosis. The interaction of antibodies and cell membranes is now being studied. These studies may show that the problem of transporting molecules in CF is due to the presence of an abnormal substance in the blood which combines with, and alters, the cell membranes.67

One of the main research topics currently is the detection of CF heterozygotes (carriers).

A pediatrician from Duke University in North Carolina, discovered that when blood samples from parents of CF children are placed on tissues taken from the tracheas of rabbits, the cilia in the tissue lose their normal, coordinated wavelike motion and begin to beat erratically. This "rabbit test" may eventually be a method used in genetic counseling. There is also a possibility that by identifying the blood faction which causes the ciliary disruption, the basic cause of cystic fibrosis could be found.68


67 Ibid., p. 2.

One of the most promising methods of CF heterozygote detection is checking meconium for albumin concentration. A newborn infant's first meconium stool is taken for the specimen. A small amount of meconium is spread on the test-strip. The test-strip is then placed immediately in a plastic vial containing three to five drops of de-ionized water. After ten minutes an intense blue coloration occurs by ascending chromotography. This only occurs if there is 20 milligrams albumin per gram dry weight of meconium. Faint blue colorations must be regarded as negative. If the test is positive, the pilocarpine (sweat) test should be run for confirmation. The major advantage of this detection method is that CF can be detected and genetic counseling can be done before another pregnancy occurs. Also, the cost is approximately forty cents per kit (test-strip/vial) which is inexpensive enough to make it feasible for all people. In the two years of testing this method, only four false negative tests have been reported.\textsuperscript{69} False positive tests can occur if blood contaminates the meconium or if the baby is premature. Prematurity results in low pancreatic function thus causing a high albumin reading.\textsuperscript{70}

Dr. H. Schwachman has found a possible alternative detecting CF in the cases where the meconium specimen has been


contaminated or if the baby is premature. The stool is checked for lactase. Lactase is seen in CF babies but not in normal ones.

\[
\text{Lactase} \\
\downarrow \\
\text{Lactose} \rightarrow \text{glucose and galactose}
\]

To test for lactase, lactose is added to the meconium stool of an infant. If lactase is present, glucose is liberated. This glucose can be detected by Dextrostix. If a positive test occurs, it can be checked by adding distilled water to the meconium rather than lactose. If the Dextrostix still has a positive result, the blue color is due to free glucose rather than lactose breakdown. 71

Another method of detecting CF carriers is presently being researched. It consists of measuring the electrical properties of rat jejunum in the presence of the blood serum being tested. The electrical activity goes up when CF heterozygote serum is used. 72

Malabsorption of fat is a characteristic feature in 90 percent of cystic fibrosis patients. Some of the blood levels of essential fatty acids are low in patients, whereas other essential fatty acids are present in excessive amounts. In the last two years, investigators have been able to measure levels of substances called prostaglandins that are similar


to hormones and exert an effect on many vital functions of certain glands such as the pancreas and the antibody system. Prostaglandins are composed of fats and are derived from the precursor fatty acids which are low in cystic fibrosis. Dr. Robert Elliot, a pediatrician in New Zealand, decided to try the effects of increasing the levels of essential fatty acids present in the blood of CF patients. In 1975, he reported his initial findings in an infant with cystic fibrosis who had been born with meconium ileus. He gave the fat to the child intravenously every few weeks. His findings were: (1) the sweat salt excretion was reduced, (2) the pancreatic function improved and (3) the child was free from lung infection. During the next two years, he tried the same thing with seven other children. He found that the younger children responded much better to this therapy than the older children. Dr. Elliot's research has been criticized, however, because he did not use a control group. At the present time Children's Memorial Hospital in Chicago is commencing a trial of fat supplements by mouth. 73

With the varied types of research being done, it is possible that the cure may be found in the near future.

Chapter VI

CYSTIC FIBROSIS FOUNDATION
Twenty-two years ago, in 1955, a small group of parents with cystic fibrotic children decided to join their scattered efforts of dealing with cystic fibrosis into a chartered, non-profit organization. In the first year, less than $100,000 was raised. Education about CF was only by word of mouth. Slowly, however, cystic fibrosis became a concern for others outside the boundaries of personal involvement. From these small starts came the National Cystic Fibrosis Research Foundation, now called the Cystic Fibrosis Foundation.

Studies aimed at clarifying the symptoms and complications of cystic fibrosis were of primary concern during the Foundation's first ten years. "The goal was to lessen the harshness of CF and to extend and normalize the lives of those affected." Presently, more emphasis is being placed upon research on how the disease affects the cells and their components.

The National Cystic Fibrosis Foundation has chapters in every state. Indiana's chapter is located at 3843 North Meridian, Indianapolis. The main function of these state chapters is fund-raising. The state director coordinates

fund raising campaigns all over the state. From the monies received, 55 percent must go the National Cystic Fibrosis Foundation. This is a national rule. Each state chapter is self-sufficient. It must run on the 45 percent of the collected funds. This money is used to pay salaries, rent or mortgage payments on the office, and extra expenses. No money collected by the state can be distributed by the state. It must go through the National Cystic Fibrosis Foundation. 75

The National Cystic Fibrosis Foundation controls the funds collected by the states. These monies are divided into two major categories--program and supporting services. See chart on page that follows.

The supporting services refers to management, general costs and fund-raising. This would include such things as printing of pamphlets, cannisters for stores, mailing, paperwork, etc.

The programs offered by the Cystic Fibrosis Foundation can be categorized under three main topics: research, education, and service. Some of the programs deal with more than one topic. See figure on page 49.

The Cystic Fibrosis Center program is heralded as one of the best developed, non-federally funded regionalized systems of care for patients with a single disease or category of

75Mr. ______________________, interview with Director of Indiana's Cystic Fibrosis Foundation, Indianapolis, Indiana, October 18, 1977.
Figure 6  Where Does Cystic Fibrosis Foundation Money Go? 76

Program ($6,542,874)
Research, Medical Care 53%
Clinical Fellowships 4%
Public and Professional Information and Education 16%
Community Services 10%

Supporting Services ($2,164,926)
Management and General 8.5%
Fund-Raising 8.5%

81% Program
8.5% Fund Raising
8.5% Management

### Figure 7  CYSTIC FIBROSIS FOUNDATION PROGRAMS

<table>
<thead>
<tr>
<th>Program</th>
<th>Research</th>
<th>Education</th>
<th>Service</th>
</tr>
</thead>
<tbody>
<tr>
<td>CF Center Program</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Young Adult Program</td>
<td>X</td>
<td></td>
<td>X</td>
</tr>
<tr>
<td>Professional Education Program</td>
<td></td>
<td>X</td>
<td></td>
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<tr>
<td>Professional Training Program</td>
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<td>X</td>
<td></td>
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<tr>
<td>Government Relations Program</td>
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<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Health Education</td>
<td>X</td>
<td></td>
<td>X</td>
</tr>
</tbody>
</table>

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diseases.\textsuperscript{78} Today there are over 100 CF Centers. The CF Centers are partially financed by the National Cystic Fibrosis Foundation. Clinics are found in either hospital or doctor's office settings. They provide expert care for children with cystic fibrosis. Besides providing care for CF children and adults, the CF Center program is also the base for many medical and scientific activities of the CF Foundation. The physicians and staff in these centers also become educators. They educate not only the family and the child but also other members of the health team. CF personnel present classes and seminars on various aspects of cystic fibrosis.

A registry of all patients is kept by the Center so that trends in the diagnosis and life span of CF patient can be analyzed. An example of this is that in 1966, about seven percent of CF Center patients were age 18 or above. At the end of 1974, almost 16 percent of CF Center patients were in this age category.\textsuperscript{79}

Indiana has four CF Centers: Riley Hospital in Indianapolis; Methodist Hospital, also in Indianapolis; Elkhart General Hospital Deaconess Hospital in Elkhart; and Evansville Hospital in Evansville. Although there are cooperative ties between the Indiana State CF Foundation and the Clinics, there are no financial ties.

The Young Adult Program was instituted in 1972. The program deals with problems such as vocational or life adjust-

\textsuperscript{78} Ibid., p. 4.

\textsuperscript{79} Ibid.