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"Review & Update: Cystic Fibrosis"

September 2017

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Cystic fibrosis (CF) is a chronic, life-threatening autosomal, recessive (genetic) disorder that is inherited from both parents whose genes have undergone genetic changes (mutations) in the Cystic Fibrosis Membrane Conductance Regulator (CFTR) protein which is encoded by the CFTR gene. This is one of a number of topics that we review every few years.

This lesson provides 1.25 (0.125 CEUs) contact hours of credit, and is intended for pharmacists & technicians in all practice settings. **The program ID # for this lesson is 0798-0000-18-235-H01-P for pharmacists, and 0798-0000-18-235-H01-T for technicians.**

Participants completing this lesson by August 31, 2020 may receive full credit. Release date for this lesson is September 1, 2017.

To obtain continuing education credit for this lesson, you must answer the questions on the quiz (70% correct required), and return the quiz. Should you score less than 70%, you will be asked to repeat the quiz. Computerized records are maintained for each participant.

If you have any comments, suggestions or questions, contact us at the above address, or call 1-843-488-5550. **Please write your name, NABP eProfile (CPE Monitor®) ID Number & birthdate (MM/DD) in the indicated space on the quiz page.**

The objectives of this lesson are such that upon completion participants will be able to:

For Pharmacists:

1. Define cystic fibrosis and relate its genetic pathogenesis.
2. Differentiate between classic and non-classic cystic fibrosis.
3. Describe the constituents of mucus and its effect on the lungs, pancreas, skin and male reproductive organs.
4. Discuss the symptoms of cystic fibrosis.
5. Describe therapeutic options.
6. List medications used.

For Technicians:

1. Define cystic fibrosis.
2. Differentiate between classic and non-classic cystic fibrosis.
3. Discuss the symptoms of cystic fibrosis.
4. List the medications used.

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CYSTIC FIBROSIS

Cystic fibrosis (CF) is a chronic, life-threatening autosomal, recessive (genetic) disorder which is inherited from both parents whose genes have undergone genetic changes (mutations) in the Cystic Fibrosis Membrane Conductance Regulator (CFTR) protein which is encoded by the CFTR gene. Mutation means changes, alteration, modification, transformation, deletion, or shortenings in a gene that may be transmitted to future generations. CF is mostly present at birth and affects cells of any glands found in the body that produce mucus, sweat, digestive fluid, and male fertility fluids. The secretions are normally watery in nature. However, in CF, such fluids gradually become thick, sticky and viscous with glue-like consistency. These fluids form plugs that obstruct ducts of the aforementioned secretory glands. Organs most critically affected by CFTR are the lungs, but the pancreas, liver, kidneys, intestines, and male reproductive system may be involved.

The name cystic fibrosis was derived from the characteristics of the disorder. The word fibrosis is derived from scar, and the word cystic is derived from the word cyst (a membranous sac containing fluid) which was first discovered in the pancreas in the last century. Currently there are about 30,000 cases of CF in the US, 75% of which are diagnosed around the age of two. About 1000 patients are diagnosed annually. Moreover, about 4% of the cases are misdiagnosed and others are diagnosed belatedly. While CF occurs in all races and ethnic groups, the incidence varies from one group to another. It is encountered more among Caucasians of North European decent, followed by Hispanics, Native Americans, African Americans and Asian Americans. The following is the frequency among different races: 1:3,000 Caucasians, 1:9,200 Hispanics, 1:10,900 Native Americans, and 1:30,000 Asian American. It is believed that these estimates may increase due to newborn screening and better methods of diagnosis. That being said, the life expectancy of CF patients has improved. The Cystic Fibrosis Foundation (CFF) Registry states that about half the patients are now older than 18 years of age and the median age of survival is 33.4 years, with patients living in their fifties and sixties compared to three decades ago where many children did not reach adulthood.

CAUSES

CF occurs as a result of a defect (mutation) in the gene CFTR which is a membranous protein, and chloride channels encoded by the CFTR gene. A gene on chromosome 7 leads to the production of CFTR. When mutation in the gene occurs, this results in the production of a defective CFTR leading to the development of CF. The mutation occurs when three nucleotides deletion takes place resulting in loss of the amino acid phenylalanine (F) at the 508th position of the protein. Genetic deletion is denoted by the sign Δ (delta). Deletion of nucleotides results in loss of part of the chromosome or a sequence of the DNA molecule with all or part of enomic material. ΔF508, delta 508, is a specific mutation within the gene for CFTR.

The CFTR gene was recognized about 25 years ago as the main gene that plays a major role in the development of CF. CFTR is a membrane protein and chloride channels which is encoded by CFTR gene in the epithelial tissues. The CFTR manufacture the CFTR protein which controls the chloride ion present within the epithelial cells found in the mucosa of the lungs, pancreas, nasal cavity, GI tract, and male reproductive parts. The cells normally secrete a thin, watery fluid such as sweat, mucous, tears, nasal, digestive and male reproductive fluids. Any delay or disruption in the migration of chloride ions from the cell through the CFTR protein will result in water retention

within the cells due to osmosis. This leads to an increase in the viscosity of those fluids. Normally proper control of chloride channels results in correct balance of electrolytes on both sides of the cell as well as maintenance of the consistency of the secretions of the involved organs. Mutation of the CFTR gene leads to disruption of regulation of cell electrolyte and water. A function of CFTR gene controls the energy transfer that causes the cell to open and close its channels. The CFTR gene which is located in chromosome 7 is composed of 27 sequences of DNA that encode 1480 amino acids.

CF is an autosomal (chromosome other than sex chromosomes, allosomes), recessive (inherited characteristics which stem from the gene) disorder with two copies of abnormal genes present. The disorder is acquired when a child inherits one mutated copy of CFTR gene from each parent. However, if the child receives only one mutated copy then the child is CF free but is considered a carrier. When a child inherits two mutated CFTR genes the child will have CF. The probability of parents who are genetic CF carriers to have a child with CF is 25%, i.e. 1 in every 4 babies. The probability of genetic CF carrier parents to have a CF carrier child is 1 in every 2 babies or 50% and 1 in every 4 children (25%) will neither have CF nor carry CF. Each child whose parents are carriers has 25% risk of inheriting 2 normal genes and 50% probability of not being a carrier. Failure of the mutated CFTR gene to regulate and control the electrolyte in the cells results in increased thickness and stickiness of secretions in the lungs, pancreas, digestive tract, skin and male reproductive system.

There are more than 1800 CFTR gene mutants. About 70% of CF cases are due to the mutants that result from deletion of one amino acid at the position 508 in the CFTR gene. Other mutants to the CFTR gene include structural changes in the protein. Stability or production can interfere with chloride ion regulations in the epithelial cells of the affected organ. Mutations other than ΔF508 have been identified, though they are rare. Mutated CFTR protein cause cells to produce excessively salty, viscous and thick fluids which accumulate in the lung ducts as well as those in the pancreas, digestive system and male reproductive system.

MUCUS

Mucus is a slippery, normal, protective, relatively viscous substance that consists of mucine (glycoprotein), electrolytes, epithelial cells, leukocytes and water. It is normally produced by mucous glands to lubricate various tissues such as the lining of the mouth, nasal cavity, lungs, gastrointestinal tract, urogenital tract, and other body cavities. It prevents drying out of the mucous membranes that it covers by keeping them moist. It also traps foreign substances such as dust, smoke, microorganisms and pollen into the nasal cavity, lungs and eyes. Mucus contains antiseptic enzymes such as lysozymes and immunoglobulin to help fight infections.

CYSTIC FIBROSIS TYPES

CF occurs in two main types:

- Classic or typical, or
- Non-classic or atypical

Classic CF is identified as such if the patient exhibits symptoms, as described earlier, in one or more of the following systems: respiratory mainly the lungs, digestive including the pancreas, urogenital and skin. Moreover, the patient experiences increased salty sweat chloride (62 mmol/L or more) which is the concentration of chloride excreted in sweat. The level of sweat

chloride is highly elevated in patients with classic CF. As a result the sweat chloride level is used to screen CF.

For infants of 6 months of age or less the diagnostic range for sweat chloride level is as follows:

- Equal to or less (≤) than 29 mmol/L indicates presence of CF is very unlikely.
- A level of 30-69 mmol/L is intermediate and indicates the possibility of the presence of CF.
- Equal to or greater (≥) than 60 mmol/L indicates the likelihood of the presence of CF.

For persons 6 months of age and older, the diagnostic concentration of sweat chloride is as follows:

- Equal or less (≤) than 30 mmol/L indicates that CF is very likely.
- Concentration of 40-59 mmol/L is intermediate and indicates the possibility of the presence of CF.
- Concentration equal to or greater (≥) than 60 mmol/L indicates the likelihood of the presence of CF.

Nonclassic CF is exhibited when a patient meets the diagnostic criteria for classic CF with one exception: patients with nonclassic CF experience normal or intermediate sweat chloride level. That being said, there is a range in the severity of CF that makes it difficult to differentiate between the two types. Nonclassic CF has similar causes, symptoms and diagnosis to classic CF, but it is a much milder form. Unlike classic CF where the symptoms are pronounced and may cover multiple organs, nonclassic CF involves one organ system with milder symptoms, in particular sweat chloride level, which may or may not be present. Nonclassic CF symptoms may fall, rise or diversify during the course of the disorder. Nonclassic symptoms may be obscure during childhood but may be diagnosed when the patient reaches adulthood. Normally life expectancy of patients with nonclassic CF is longer than those with classic CF and quality of life is better.

SYMPTOMS OF CF

Symptoms of CF vary from person to person in frequency, severity and fluctuation. The symptoms may improve or intensify at times. The earliest sign of classic CF occurs when the child's sweat is observed to be salty as for example when the mother kisses her baby. Moreover, the infant experiences difficulty in passing the earliest stools after birth. The earliest stool passed by infants is known as meconium, and consists of ingested substances when the infant is in the womb—epithelial cells from the intestines, soft unpigmented hair that may be found on the body of the fetus or newborn infants known as lanugo, mucus, protective amniotic fluid sometimes known as pregnant woman water and is present in the amniotic sac, and bile. Some patients may experience symptoms only when they reach adolescence or adulthood. The following are the signs and symptoms encountered in various organ systems. The major ones affect the respiratory system, specifically the lungs, followed by the digestive and the male reproductive system.

The main symptoms that occur in the lungs is the production of sticky, viscous and thick mucus that sometimes may be mixed with blood. This mucus bonds with the lining of the bronchial airways forming obstructive plugs that trigger persistent productive cough, wheezing, breathlessness (dyspnea) as well as expectoration of glue-like phlegm or sputum.

The mucus is a good medium for the multiplication of microorganisms. As a result, the patient becomes vulnerable to frequent episodes of respiratory infections which may be caused by *Streptococcus pneumonia, Staphylococcus aureus, Hemophillus influenza, and Psuedomonas aeruginosa*. Patients suffering from CF are more prone to respiratory infections caused by mucoid Pseudomonas than healthy persons free of CF.

Repeated lung infections as well as the presence of inflammation within the bronchial airways and obstructive mucus can lead to structural changes within the lungs, which lead to more complications and vulnerability to infection, breathlessness and weakness. Diminished airway clearance of mucus may occur due to damage to the cilia present on the trachial and bronchial epithelium surfaces. In later stages, the damage and the anatomical changes that occur in the lungs cause the airways to become permanently enlarged (bronchiectasis). Such complication may result in immobility to expel mucus and in deterioration of airway structure causing exacerbation of breathing difficulties. In the presence of bronchiectasis, production of mucus may reach 240 ml daily. Patients with dry bronchiectasis may cough blood in the absence of phlegm. Sinusitis, bronchitis and nasal polyps, which may interfere with breathing are frequently encountered.

The nasal cavity secretions are normally watery but their consistency may become viscous and may clog the sinus passages leading to infection, facial pain and headache.

Mucus formation in the digestive tract can block the tubes and ducts of the pancreas thereby preventing pancreatic enzymes that digest fat and protein from reaching the intestines through the duodenum. Lack or reduced fat absorption leads to accumulation of unabsorbed fat in the intestinal contents resulting in steatorrhea (excretion of higher than normal quantities of fat in the feces, which is usually bulky, oily in appearance and foul smelling and may lead to an oily anal leakage and fecal incontinence). Blockage of the intestines may occur especially in the newborn. Lack of nutrient absorption may result in malnutrition and suppression of growth in children as well as hypoproteinemia (characterized by generalized edema), inability to fully absorb fat soluble vitamins A, D, and K.

Pancreatic obstruction by thick mucus may cause pancreatitis, fibrosis of the pancreas and atrophy of the exocrine glands in case of complete shutdown of the ducts, thereby cutting off the insulin produced by the islet of Langerhans cells. Rectal prolapse (slippage of the lower part of the large intestine outside of the anus) may occur. Bile duct obstruction may lead to cirrhosis, hepatitis and jaundice due to increased blood level of bilirubin. Liver failure may lead to accumulation of toxins in the blood and deficiency in the protein that plays an important role in blood clotting. Bile ducts that normally carry bile from the liver to the gall bladder and small intestine where it assists in fat digestion, may fail. Other digestive tract problems that may be created by the presence of the mucus is the folding of one part of the intestine within another leading to a serious intestinal obstruction resulting from increased feces thickness. Constipation as well as heartburn may be encountered.

The reproductive system in males may be affected as CF can retard the development of the vas deferens (duct that delivers sperms from the testicles to the urethra) causing infertility but not sterility. Men with such a defect produce sperms and enjoy coitus but can conceive only artificially through assisted reproductive technology (ART). Sperms are stored in the testicles and in the epididymis until ejaculation. Absence or blocked vas deferens will prevent sperms from reaching the semen. Older men with CF have been discovered to have had undiagnosed

mild CF which led to absence of the vas deferens. About 90% of men with CF are infertile, while about 20% of women with CF are infertile due to thickened cervical mucosa. Thus, the majority of women can become pregnant since CF does not affect the development of the female reproductive system. However, the thick mucus can interfere with fertilization of the egg.

Sweat glands that produce salty tasting sweat on the skin are affected by CF. Depletion of salt especially on hot days or after performing strenuous exercises or during fever could lead to fatigue, weakness, muscle cramps, vomiting, dehydration and heat or sun strokes, especially in persons 50 years or older.

DIAGNOSIS

As indicated earlier the skin of CF patients is covered with salty sweat. Thus the presence of a salty-tasting skin is utilized as a diagnostic measure for CF. Other clinical signs which are encountered include growth suppression in children due to malabsorption of nutrients from the intestinal tract, weight and height loss, increasing quantity (over-accumulation) of sticky mucus, repeated respiratory infections, productive cough, dyspnea, male infertility, bowel obstruction mainly during infancy. Beside the aforementioned clinical signs, CF can be diagnosed during the first month of life through screening of the newborn. Screening methods include high level of immunoreactive trypsinogen (IRT) in the infant's blood, which is released by the pancreas. This test by itself is not reliable as a high IRT level may occur following premature birth or difficult delivery. To assist in the confirmation of CF, genetic tests may be performed to determine the presence of gene defects in infants as young as a few weeks old.

CURRENT THERAPY

There is no known cure for CF. However, there are a number of therapeutic measures that can be taken to reduce the severity of the symptoms, lessen the occurrence of complications, improve the quality of life, and prolong life expectancy. The earlier the diagnosis and intervention, the better chance of successful management of CF. As indicated earlier, the severity of the symptoms vary from one person to the next. The severity of the symptoms may fluctuate from time to time. As a result, treatment should be individualized to each patient.

The goals of therapy include:

1. Preventing and combatting infections, especially the respiratory ones.
2. Assisting in airway clearance by liquification and expectoration of accumulated mucus from the throat and lungs.
3. Treating and preventing internal obstruction such as nasal polyps and intestinal blockade.
4. Availing proper quality and quantity of nutritious foods and fluids to prevent malnutrition and dehydration, which may result in thicker mucus.
5. Improving the quality of life and boosting organ function by physical and chest therapy.

Treatment Options

1. **Medications:** The administration of pneumococcal vaccine can help in providing resistance to bacterial lung infections. Antibiotics should be used in both the prevention and treatment of lung infections.

2. **Airway clearance** can be improved by following several techniques. Parents should lend assistance to infants and toddlers to achieve airway clearance. Older children and adults can select the method that is comfortable and useful for them. A respiratory therapist may explain the methods used and the patient may try each method and with the recommendation of the therapist may decide on following a convenient and effective technique. Coughing can occur without control of the patient as a result of involuntary reflex initiated by the presence of mucus in the airways. Also, coughing can be initiated by the patient to clear the bronchial airways. Mucus in the large tubes of the bronchial tree can be expectorated by coughing, whereas mucus in the small and narrower branches can only be moved to larger tubes by coughing after which it may be expelled to the outside. Tapping the surface of the chest by using light rapid blows may assist in loosening the mucus. The use of inhalation containing bronchodilators that assist in relaxing or reducing inflammation of the smooth muscles around the airways is recommended.
3. **Nasal polyps** which may interfere with breathing, need to be removed.
4. **Blockages of the tubes in the pancreas can prevent the digestive pancreatic enzymes from reaching the intestine resulting in malnutrition, bulky stools, flatulence, severe constipation, discomfort and pain.**
5. **The intake of nutritious food is important for good health in general, and the CF patient in particular.** Gaining adequate weight may improve lung function. Some patients may require feeding tubes to insure that they are receiving adequate nutrients. Such tubes may be placed surgically in the stomach or through the nose by means of a nasal gastric tube.
6. **Improving the quality of life through support** of the family members, maintaining nutritious diet after consultation with an experienced dietitian, maintaining the patient's weight or increasing it improves lung function.
7. **Chest physical and oxygen therapies** should be maintained in case there is a need for these.

The goal of **chest physical therapy** is to loosen the sticky mucus that is normally present in the lungs. One method calls for clapping with cupped hands on both sides of the chest, 4 times daily. A specially designed tube in which the patient breathes is used. A chest vibrator may be used. Pulmonary rehabilitation which includes physical exercise and breathing techniques designed by a therapist tend to enhance the lung function.

Oxygen therapy helps maintain adequate oxygen level in the blood. Low blood oxygen can result in elevation of blood pressure within the lungs (pulmonary hypertension) as well as breathing difficulty, tiredness, and confusion. Oxygen therapy provides the patient with additional oxygen to breathe which is delivered through devices available in various forms, types and sizes depending on the intended use. It may be delivered from a tube placed in the nose or from a facemask. Some are portable or mobile and more convenient than the home oxygen. Portable oxygen concentrators work by removing nitrogen from the air leaving a high oxygen concentration in the breathed air. The oxygen can be stored as a gas or liquid in specially designed tanks that can be refilled. Oxygen therapy can be used for short or long durations in both the hospital, at home or during the performance of daily functions. Patients and their families need to be aware of the fact that oxygen present in the devices is a fire risk, thereby the use of flammable materials should be avoided when using the devices. Even though oxygen therapy is safe, side effects such as dry nose and tiredness may occur.

NEW THERAPY

Ivacaftor is used for treating CF caused by certain mutants of CFTR and in combination with lumacaftor for CF caused by ΔF508 mutation. The drug is aimed at treating the cause rather than the symptoms of CF. Ivacaftor was approved by the FDA in January 2012, but the combination therapy was approved in July 2015 for patients 12 years of age and older who have mutation ΔF508. Ivacaftor is used by itself in patients who have one of the following mutations in CFTR protein: G551D, G1244E, G1349D, G178R, G5515, 512512N, 51255P, 5549N, 5549R or R117H. Side effects of ivacaftor include abdominal cramps, diarrhea,

dizziness, upper respiratory infections, dyspnea, nasal congestion, nasopharyngitis, and headache. Ivacaftor acts by enhancing the opening of chloride channels, thereby allowing the chloride ion to pass through. The drug should not be taken with grapefruit juice, grapefruit or Seville Oranges (bitter orange, sour orange). This orange is a native to Southeast Asia and can be found in Florida and the Bahamas.

MEDICATIONS USED IN THE TREATMENT OF CF SYMPTOMS

Mucolytics are drugs that act by liquefying the thick mucus that bonded to the surface of the bronchial airways. The liquification process occurs by breaking up the chemical bonds of the mucosa. N-acetylcholine has been used as a mucolytic, but its effectiveness has not been proven. Dornase alfa has been shown to improve pulmonary exacerbation.

Expectorants tend to increase bronchial glands secretions resulting in an increase in the level of hydrolysis of mucin. Sodium and potassium citrate, as well as potassium iodide and guaiphenesin may be used.

Antiinflammatory drugs such as the corticosteroids may be used to reduce inflammation and swelling in the airways of the lungs. This can be administered via inhalations.

Bronchodilators are used as inhaled medications to relax the smooth muscles around the bronchial tube thereby improving inhalation and exhalation.

Pancreatic enzymes are used to assist in digestion of fats and other nutrients in the diet. These orally administered medications may be combined with vitamin supplements. Lipase acts with bile from the liver to help digest fat; protease tend to break down protein; amylase digests carbohydrates to more easily absorbed sugars.

SUMMARY

Cystic fibrosis is a chronic, life-threatening autosomal recessive disease which is inherited from both parents whose genes have undergone genetic changes in the CFTR. It most commonly affects the lungs, but the pancreas and intestine are prone to this disorder. The hallmark of cystic fibrosis is the production of sticky, glue-like mucus within the bronchial airways thereby causing breathing difficulties, coughing, wheezing, and fatigue. Cystic fibrosis of the pancreas can prevent pancreatic enzymes from reaching the intestines causing inadequate digestion of fat, protein and carbohydrates which leads to malabsorption. There are about 30,000 cases of CF in the U.S., most of which are children and young adults. Due to newborn screening and better methods of diagnosis, the life expectancy of CF patients has improved. About half of patients are now older than 18 years of age and the median age of survival is 33.4 years. There is no known cure for CF. However, there are a number of measures that can be taken to reduce the severity of symptoms.

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Contributing Faculty/Authors
Farid Sadik, PhD
Dean Emeritus University of South Carolina
College of Pharmacy
Columbia, SC
Adjunct Professor
Presbyterian College
School of Pharmacy

Executive Editor
William J. Feinberg, RPh, MBA

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LESSON EVALUATION

Please fill out this section as a means of evaluating this lesson. The information will aid us in improving future efforts. Either circle the appropriate evaluation answer, or rate the item from 1 to 7 (1 is the lowest rating; 7 is the highest).

1. Does the program meet the learning objectives?
   - Define CF — YES NO
   - Compare classic & nonclassic CF — YES NO
   - Discuss symptoms of CF — YES NO
   - Describe therapies associated with CF — YES NO

2. Was the program independent & non-commercial? YES NO

	Low Relevance						Very Relevant
3. Relevance of topic	1	2	3	4	5	6	7

4. What did you like most about this lesson?_____

5. What did you like least about this lesson?_____

Please Mark the Correct Answer(s)

1. **Cystic fibrosis occurs as a result of:**
   A. Infection acquired immediately after birth.
   B. An autoimmunity reaction.
   C. Inheritance from parents whose genes have undergone CFTR mutations.
   D. Inheritance from one parent.

2. **Epidemiological studies indicate that CF is more common in:**
   A. Caucasians B. Hispanics
   C. African Americans D. Asian Americans

3. **Most CF cases occur due to deletion of one amino acid at position _____ in the CFTR?**
   A. 805 B. 611
   C. 511 D. 508

4. **A diagnostic feature in CF is:**
   A. Sweat chloride concentration less than 30 mmol/L.
   B. Increased salty sweat chloride at concentration of 60 mmol/L or more.
   C. Normal concentration of sweat chloride.
   D. Salty taste of skin after age 6 years.

5. **Which statement is true regarding portable oxygen concentrators?**
   A. Use only liquid oxygen.
   B. Have no side effects.
   C. Used only for children.
   D. Deliver oxygen to the lungs by removing nitrogen from the air

6. **CF affecting the pancreas could lead to:**
   A. Hepatitis C. B. Cutting off insulin.
   C. Kidney stones. D. Over production of amylase.

7. **What is a symptom of CF in a male adult?**
   A. Loss of prostate fluid.
   B. Dead sperm.
   C. Sterility.
   D. Infertility.

8. **Which statement is true about ivacaftor?**
   A. Used to treat people with G551D mutations in CFTR.
   B. Was approved by FDA in 2008.
   C. Only used to treat symptoms of CF.
   D. To be effective for ΔF508 it must be taken alone.

9. **Which statement is true about ivacaftor?**
   A. Shuts off chloride channels.
   B. Used with lumacaftar for patients with CF who have mutation other than ΔF508.
   C. Do not take with grapefruit juice.
   D. Used only in children 4 years of age or younger.

10. **Which of these is true?**
    A. Life expectancy in classic CF is longer than in nonclassic CF.
    B. Life expectancy in classic CF is shorter than in nonclassic CF.
    C. Life expectancy in classic CF equals that of nonclassic CF.
    D. None of these.