

Cystic Fibrosis

Background:

- ❖ “Cystic fibrosis (CF) is an inherited chronic disease that affects the lungs and digestive system of about 30,000 children and adults in the United States (70,000 worldwide).” (CF Foundation, 2007)
- ❖ A chronic disease unfortunately is a disease in which is on going and never comes to an end until it takes the individual’s life away.
- ❖ This chronic disease affects a gene in the body in which a protein is produced that makes the body make mucus that is very thick and sticky. With one’s body producing very thick and sticky mucus it will start to block the lungs and this causes an infection in the lungs, which eventually becomes life-threatening. Not only does it affect the lungs but also affects the pancreas. In the pancreas, it will not allow natural enzymes to form, which causes a problem because now the body is not able to absorb any food or even break it down. (CF Foundation, 2007)
- ❖ Signs of CF are when one is sweating, they produce a lot more salt, lung problems and infections, constant cough, always being short of breath and wheezing with very little exercise involved, abnormal weight gain, and having a trouble going to the bathroom. (CF Foundation, 2007)
- ❖ The treatment for CF is therapies and lung transplants. Patients must undergo regular routines to keep the mucus thin because if not, then it’s going to get to the point where it’s going to block up the lungs and the patient will not be able to breath and this is when the disease takes the patient’s life away. (CF Foundation, 2007)

Statement of the Problem:

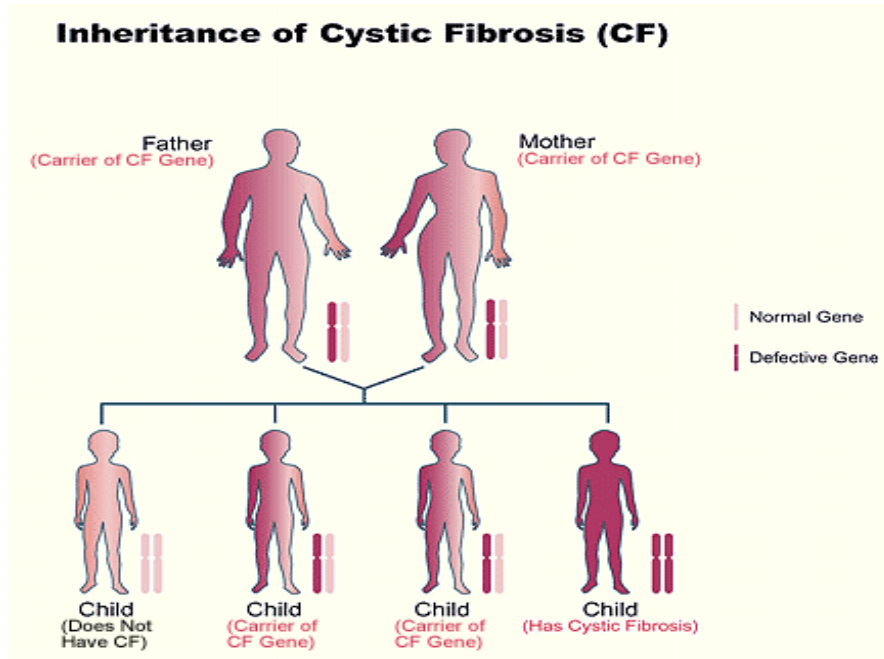
- ❖ “Cystic fibrosis (CF) is an inherited disease of your mucus and sweat glands. It affects mostly your lungs, pancreas, liver, intestines, sinuses, and sex organs.” (Health & Human Services, 2007)
- ❖ Cystic Fibrosis happens when a genetic mutation ends the making of new proteins in the cells of the lungs, pancreas and other organs. Having no protein damages the cells that transport chloride ions into and out of the cell. This causes the buildup of mucus, which then leads to buildup of bacteria and then causes lung infections. (Cystic Fibrosis, What is it, 2008)
- ❖ “Tests to diagnose cystic fibrosis can be done at any time-before pregnancy, during pregnancy, in childhood, or in adulthood. Genetic tests for couples who are planning a pregnancy or who are expecting a baby can help determine whether either person is a carrier of the gene that causes cystic fibrosis.” (Genetics Home, 2008)
- ❖ Treatment for CF is through therapies and lung transplants. Between these two, the individual has a chance to live longer than expected. (Health & Human Services, 2007)
- ❖ Goals for CF is to continually increase the life expectancy and hopefully find a cure for it, although it will be very hard because it is dealing with genes and it's hard to stop the body from doing something genetically. (Cystic Fibrosis, What is it, 2008)

- ❖ Between the treatments and lung transplants, they are on their way of figuring out what exactly is going on and want to continue to research what is going on with the genes that are causing these mutations. There is a list of ideas on how to prevent this, a few are eat high calorie foods, avoid second hand smoke, and undergo frequent tests and check ups so doctors can catch it early. (Cystic Fibrosis, What is it, 2008)

Epidemiologic Picture:

- ❖ “The life expectancy for people with cystic fibrosis has been steadily increasing over the past 40 years. On average, people who have cystic fibrosis live into their mid-to-late 30s, although new treatments are making it possible for some people to live into their 40s and longer.”
- ❖ Males and females are capable of getting Cystic Fibrosis. (Health & Human Services, 2007)
- ❖ Cystic Fibrosis is generally found in Caucasians. (Health & Human Services, 2007)
- ❖ “About 1,000 new cases of cystic fibrosis are diagnosed each year.” (CF Foundation, 2007)
- ❖ “More than 70% of patients are diagnosed by age two.” (CF Foundation, 2007)
- ❖ “More than 40% of the CF patient population is age 18 or older.” (CF Foundation, 2007)
- ❖ “In 2006, the predicted median age of survival was 37 years.” (CF Foundation, 2007)

- ❖ In the United States there are about 30,000 children and adults with CF and about 70,000 in the world with Cystic Fibrosis. (Health & Human Services, 2007)
- ❖ 1 out of 3,000 babies born are diagnosed with CF. (Health & Human Services, 2007)



(Health & Human Services, 2007)

Solutions:

Genetics Home Reference (<http://ghr.nlm.nih.gov/condition=cysticfibrosis>)

- ❖ Genetic Home Reference has recently been reviewed in January 2008, providing readers with additional information regarding Cystic Fibrosis. This Web site is very helpful for those who are not too sure on medical terms because it breaks all the information down to the simplest forms for readers and provides other Web sites that further explains the issues and problems with CF. The issue is being

addressed in the break down from what it is, to how common it is, genes involved and the inheritance of CF.

OMIM – Cystic Fibrosis

(<http://www.ncbi.nlm.nih.gov/entrez/dispomim.cgi?id=219700>)

- ❖ 1966-2008 Johns Hopkins University provides a great deal of information regarding the issue of CF. This particular Web site discusses each of the problems that arises while having CF and explains in detail about what goes wrong and why it does what it does. The detail on this Web site is very helpful and useful for dealing with CF.

Cystic Fibrosis (<http://www.ncbi.nlm.nih.gov/books/by.fcgi?rid=gnd.section.242>)

- ❖ The NCBI has a pretty neat Web site dealing with Cystic Fibrosis because it talks about the chromosomes and explains what is going on in them. They provide diagrams to help the readers.

Internet Sources:

- ❖ National Human Genome Research Institute. (2007) *Learning about Cystic Fibrosis*. Retrieved on March 9, 2008 from <http://www.genome.gov/10001213>
- ❖ The Cystic Fibrosis Center at Stanford. (2008). *A guide to living with CF*. Retrieved on March 9, 2008 from <http://cfcenter.stanford.edu/>

- ❖ Cystic Fibrosis: University of Maryland Medical Center. 2008. *Cystic Fibrosis*
Retrieved on March 9, 2008 from <http://www.umm.edu/ency/article/000107.htm>

Bibliography:

- ❖ Cystic Fibrosis: WedMD. (2005-2008) *Cystic Fibrosis-Topic Overview*. Retrieved on March 9, 2008 from <http://children.webmd.com/tc/cystic-fibrosis-topic-overview>
- ❖ Cystic Fibrosis Foundation. (2007) *What you need to know*. Retrieved on March 9, 2008 from <http://www.cff.org/AboutCF/>
- ❖ Cystic Fibrosis: What is it? (2008). *Cystic Fibrosis*. Retrieved on March 9, 2008 from <http://www.ygyh.org/cf/cause.htm>
- ❖ What Is Cystic Fibrosis: U.S. Department of Health & Human Services. (2007) *Cystic Fibrosis* Retrieved on March 9, 2008 from http://www.nhlbi.nih.gov/health/dci/Diseases/cf/cf_what.html

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Cystic Fibrosis (Class Handout)

- “Cystic fibrosis (CF) is an inherited disease of your mucus and sweat glands. It affects mostly your lungs, pancreas, liver, intestines, sinuses, and sex organs.” (Health & Human Services 2007)
- “A defective gene and its protein product cause the body to produce unusually thick, sticky mucus that:
 - clogs the lungs and leads to life-threatening lung infections; and
 - obstructs the pancreas and stops natural enzymes from helping the body break down and absorb food.” (CF Foundation, 2007)
- **People with CF can have a variety of symptoms, including:**
 - very salty-tasting skin;
 - persistent coughing, at times with phlegm;
 - frequent lung infections;
 - wheezing or shortness of breath;
 - poor growth/weight gain in spite of a good appetite; and
 - frequent greasy, bulky stools or difficulty in bowel movements.(CF Foundation, 2007)
- **Statistics**
 - About 1,000 new cases of cystic fibrosis are diagnosed each year.
 - More than 70% of patients are diagnosed by age two.
 - More than 40% of the CF patient population is age 18 or older.
 - In 2006, the predicted median age of survival was 37 years. (CF Foundation, 2007)