

## *Cystic Fibrosis Webquest*

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### **Introduction:**

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). In the 1950s, few children with cystic fibrosis lived to attend elementary school. Today, advances in research and medical treatments have further enhanced and extended life for children and adults with CF. Many people with the disease can now expect to live into their 30s, 40s and beyond.

Cystic fibrosis is a life-threatening genetic disease that causes mucus to build up and clog some of the organs in the body, particularly the lungs and pancreas. When mucus clogs the lungs, it can make breathing very difficult. The thick mucus also causes bacteria (or germs) to get stuck in the airways, which causes inflammation (or swelling) and infections that lead to lung damage.

Additionally, the conditions in the lung of a patient with cystic fibrosis allow for colonization by a variety of microbes. These bacterial infections prove to be fatal for many patients with CF and they are treated aggressively with antibiotics throughout the course of the disease.

### **Part 1 – Basic Genetics Review**

Use your text to define the following important terms.

*“Genetics (from Ancient Greek “genitive” and “origin”), a discipline of biology, is the science of heredity and variation in living organisms.”*

### **Define:**

- Gene
- Allele
- Chromosome
- Cell
- DNA
- RNA
- Mutation
- Punnett Square
- Protein
- Amino acids
- Molecular Genetics
- Homozygous
- Heterozygous
- Genotype
- Phenotype

## Part 2 - Review of Autosomal Recessive Inheritance Pattern

Go to: <http://www.genetics.com.au/pdf/factsheets/fs08.pdf>. Answer questions and review diagram on page 2.

*“We are all carriers of a small number of autosomal recessive changes in genes that cause no symptoms.”*

1. What is a mutation?
2. Our \_\_\_\_\_, located on the \_\_\_\_\_ of our cells, provide information for the growth, development and function of our cells.
3. What are the four most common patterns of inheritance for a genetic disorder?
4. Describe how someone can be a genetic carrier for a disease, but exhibit no symptoms of that disease.

5. Fill in the Punnett Square for autosomal recessive inheritance when both parents are unaffected genetic carriers for the condition.


6. When only one of the parents is a carrier for the condition, is it possible to have an affected child? Why or why not?
7. What is the percentage chance that a child will be a carrier for the autosomal recessive disease if both of his/her parents were carriers?
8. What are the four diseases that are listed as examples of having an autosomal recessive inheritance pattern?

### Part 3 – Cystic Fibrosis: The Disease

Go to: [http://www.virtualmedicalcentre.com/humanatlas1/vmc\\_white.asp?anid=0073](http://www.virtualmedicalcentre.com/humanatlas1/vmc_white.asp?anid=0073)

Watch animation and answer questions.

1. Which three body systems does cystic fibrosis effect?
2. Which type of cells does this disease effect?
3. Name three places in the body where you can find these cells.
  - a.
  - b.
  - c.
4. The cystic fibrosis gene causes what type of defect?
5. What are the four negative effects of the sticky mucus listed in the film?
  - a.
  - b.
  - c.
  - d.
6. What are common symptoms of cystic fibrosis?
  - a.
  - b.
  - c.
  - d.

### Part 4: Cure for Cystic Fibrosis

Go to: [http://www.pbs.org/wgbh/nova/programs/ht/qt/2809\\_09.html](http://www.pbs.org/wgbh/nova/programs/ht/qt/2809_09.html). Watch video “Finding Cures is Hard” and answer questions.

*“The genome is a storybook that's been edited for a couple billion years. And you could take it to bed like A Thousand and One Arabian Nights, and read a different story in the genome every night.” -Eric Lander*

1. What did Rily Demanche test positive for?
2. What organ system does this disease primarily effect?
3. What do the victims of CF suffer from?
4. In the 80s scientists discovered the gene responsible for causing CF, the most common inherited fatal disease in this country. Instead of gene therapy, a proposed cure for CF, scientist are now looking at \_\_\_\_\_.
5. A proteins starts out as a chain of \_\_\_\_\_, but don't work as a straight line. Proteins need to be \_\_\_\_\_. Proteins can be thought of as tangles of ribbon. In the protein world your \_\_\_\_\_ is your destiny.
6. The healthy protein acts as a chute for \_\_\_\_\_ to enter and leave the cell. But if the protein is not in the right shape, it is not allowed into the membrane and \_\_\_\_\_ gets stuck in the cell and creates sticky, thick \_\_\_\_\_.
7. Why does Professor Eric Lander say that the genome project was a “piece of cake”? Why are proteins so much harder to work with?

### **Part 5: Evolutionary Questions-Why do harmful mutant alleles persist in a gene pool?**

Go to: [http://www.pbs.org/wgbh/evolution/educators/course/session7/explain\\_b\\_pop1.html](http://www.pbs.org/wgbh/evolution/educators/course/session7/explain_b_pop1.html) and answer questions

*“Because of natural selection, different alleles are more likely to confer a survival advantage in different environments. Cycles of infectious disease prevalence and virulence often reflect natural selection.”*

1. What is a balanced polymorphism?
2. What are the five diseases listed where a heterozygote genotype confers some sort of advantage?
3. What type of diseases does the anatomical defect in CF protect against?
4. How does the cholera bacteria effect individuals?

5. What do people infected with cholera usually die of?
6. What does the protein product of the CF gene control?
7. Explain why a person with the CF gene cannot contract cholera.
8. What is the selective advantage of being a CF carrier?

### Part 6 – Bacteria and Antibiotic Resistance in Cystic Fibrosis

*“Mortality in people with cystic fibrosis is due in large part to lung infections with the bacteria *Pseudomonas aeruginosa*. By age 18, about 80 percent of patients are chronically infected with this pathogen, which promotes an inflammatory response that destroys lung tissue.*

*Even with advances in antibiotic therapies — which have been responsible for increasing the median survival of patients from 14 in 1969 to 33.4 in 2001 — patients still typically lose 2 percent to 3 percent of their lung function each year.”*

Got to:

<http://www.cff.org/UploadedFiles/treatments/Therapies/Respiratory/Antibiotics/Bacteria%20and%20Antibiotic%20R%20in%20CF%20pamphlet%2010-11-05.pdf>. Answer questions

Define the following terms:

- Bacteria
- Antibiotic
- Antibiotic resistance
- Intrinsic resistance

1. What is an example of a bacterium with intrinsic resistance to antibiotics?
2. What are the two most common types of bacteria that can acquire resistance?
3. How does antibiotic resistance develop?

4. What are the three most common types of antibiotics that are used to treat infections?
5. What is a sputum culture?
6. How are the bacteria that are isolated from sputum cultures classified (5 classifications)? Describe each classification
  - a)
  - b)
  - c)
  - d)
  - e)
7. How can antibiotic resistance be prevented?
8. Based on what you have learned, why might the CF lung be an attractive environment for bacteria?