Self Help

by Kirsten Weir

A young scientist is searching for the cure to the disease that could kill him.

Thirty-year-old Jeff Carroll is fit and healthy, but he knows he won't stay that way forever. Carroll has seen the future. He has the gene for *Huntington's disease*, an incurable, fatal *neurological* (brain) disorder.

Carroll doesn't have any symptoms yet, and he's not willing to wait around for the disease to claim him. As a neuroscience researcher at the University of British Columbia (UBC), in Vancouver, he is searching for a cure.

Inherited Disorder

Huntington's strikes one in every 10,000 Americans. Symptoms begin to show up in most people between the ages of 30 and 50. The first are usually depression, forgetfulness, clumsiness, and mood swings. As time goes on, mental deterioration sets in and uncontrollable jerking movements develop. People eventually lose the abilities to walk, speak, and care for themselves.

Huntington's disease is a *genetic* disorder, passed through DNA from parent to child. Both Carroll's grandmother and mother died of the disease. As a child, he knew his grandmother was ill. "But we never really talked about what it was," he says.

Carroll was 20 when his mother began showing symptoms of the same disease. When she was given a diagnosis of Huntington's, Carroll learned that he, too, was at risk. He had a 50-50 chance of inheriting the disease. "I decided to get tested right away," he says. "I had to know." The test confirmed his worst fears.

Carroll was serving in the U.S. Army in Europe when he tested positive. The result prompted him to take Internet biology courses in his free time so he could better understand how the disease would affect his body. "I was really fascinated," he recalls. After leaving the service, he returned to school and earned a bachelor's degree in biology. He's now researching Huntington's as he completes a doctorate in neurobiology at UBC.

Bad Stutter

A *gene*, the basic unit of heredity, is made up of a long string of chemical building blocks called *bases*. Four bases spell out a person's entire genetic code. They are adenine, cytosine, guanine, and thymine, or A, C, G, and T for short.

Everyone has a gene called the *huntingtin* gene, Carroll explains. In most people, the huntingtin gene has a section in which the bases C-A-G are repeated 17 or 18 times. However, some people inherit a *mutation*, or change in their genetic code, that causes the C-A-G section to repeat itself many more times. "It's like a stutter," he says. People who inherit too many C-A-G repeats will develop Huntington's.

"Anything above 37 repeats is associated with the disease," Carroll says. The more C-A-G repeats a person has, the younger he or she will be when symptoms first show up. Carroll has 42 C-A-G repeats, so he'll probably start showing symptoms around age 49. Until that day comes, he and the other researchers in his lab are trying to understand exactly how C-A-G repeats damage the brain.

Cutting Enzyme

Every gene holds the instructions for the production of a different protein in the human body. The huntingtin gene, Carroll says, writes the code for a "big, complicated protein," called, not surprisingly, the *huntingtin protein*.

In people with Huntington's disease, the huntingtin protein is faulty, leading to the death of *neurons*, or nerve cells, in the brain. The huntingtin protein is made up of a string of molecules, similar to pearls strung on a necklace. People with Huntington's disease have too many "pearls" on that string. Scientists believe the faulty protein kills neurons when an *enzyme*, called caspase 6, slices the protein into two pieces, setting free the extra fragment of pearls to attack the neurons. An enzyme is a substance that speeds up reactions in the body.

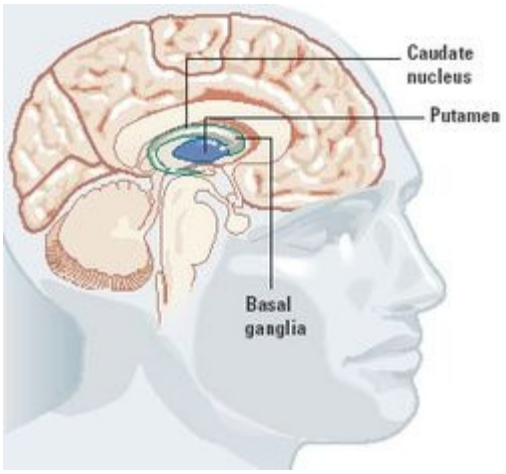
Carroll and his colleagues at UBC are working with a strain of mice engineered to carry the human gene for Huntington's. The mice exhibit symptoms of the disease. In 2006, some of Carroll's lab mates made a breakthrough. They found a way to genetically alter the strain of mice again so that the caspase 6 enzyme could not chop the faulty huntingtin protein in two. Although their huntingtin genes had more than 100 C-A-G repeats, Carroll says, "the mice didn't get Huntington's disease. They were cured."

Unfortunately, the same technique can't be applied to people. "We can't genetically engineer humans the way we do mice," Carroll says. Instead, he's looking for a drug that will protect the faulty huntingtin protein from the cutting enzyme.

Although Carroll is racing against time, he has good reason to be optimistic about finding a treatment or even a cure for Huntington's disease. The huntingtin gene wasn't discovered until 1993, and scientists have made great strides since then.

"I'm doing what I can, and that's all I can do," Carroll says. "If I didn't think it would help before my time is up, I wouldn't be here."

Loss of Nerves



Huntington's disease kills nerve cells in the *basal ganglia*, a region of the brain that controls movement and possibly *cognition* (thinking, reasoning, and remembering). The disease also affects several other areas:

- **Caudate nucleus.** This area, which is involved in motor control, shows severe shrinkage in people who have the disease.
- **Putamen.** This area is also involved in motor control and is affected in advanced cases of the disease.

The discovery of the mutant gene responsible for Huntington's has enabled doctors to predict who will get the disease and when it will begin to develop. Researchers may eventually devise a gene therapy to treat it.

Name: _____ **Date:** _____

1. What is Huntington's disease?

A. a curable, fatal neurological (brain) disorder

B. an incurable, nonfatal neurological (brain) disorder

C. an incurable, fatal neurological (brain) disorder

D. a curable, nonfatal neurological (brain) disorder

2. The text is divided into sections with subheadings. In the section called "Inherited Disorder," what does the author describe?

A. the mutation that causes the C-A-G section of the huntingtin gene to repeat more than 17 or 18 times

B. the huntingtin protein and the work that Carroll and his colleagues at UBC are conducting on mice

C. the loss of nerve cells in the region of the brain that controls movement and possibly cognition

D. the characteristics of Huntington's disease and Carroll's family history with the disease

3. Read this sentence from the text.

"Huntington's disease can have serious consequences on people."

What evidence from the text supports this statement?

A. Everyone has a gene called the huntingtin gene. In most people, the huntingtin gene has a section in which the bases C-A-G are repeated 17 or 18 times.

B. Symptoms of the disease include mental deterioration and uncontrollable jerking movements. People eventually lose the abilities to walk, speak, and care for themselves.

C. Every gene holds the instructions for the production of a different protein in the human body. The huntingtin gene writes the code for the huntingtin protein.

D. Some people inherit a mutation, or change in their genetic code, that causes the C-A-G section of the huntingtin gene to repeat itself many times.

4. In 2006, some of Carroll's lab mates made a breakthrough in their work with a strain of mice engineered to carry the human gene for Huntington's.

Why did the mice not get the disease even though they had more than 100 C-A-G repeats?

A. because the faulty huntingtin protein did not chop the caspase 6 enzyme in two and set free the extra fragments that would have attacked neurons

B. because the caspase 6 enzyme did not chop the faulty huntingtin protein in two and set free the extra fragments that would have attacked neurons

C. because the extra fragments did not chop the faulty huntingtin protein in two and set free the capase 6 enzyme that would have attacked neurons

D. because the neurons did not chop the faulty capase 6 enzyme in two and set free the faulty huntingtin protein that would have attacked extra fragments

5. What is this passage mostly about?

A. Huntington's disease and Jeff Carroll's search for a cure

B. the basal ganglia and its control over movement and cognition

C. the death of Jeff Carroll's grandmother to Huntington's disease

D. the scientific discovery of the huntingtin gene in 1993

6. Read these sentences from the text.

"Thirty-year-old Jeff Carroll is fit and healthy, but he knows he won't stay that way forever. Carroll has seen the future. He has the gene for Huntington's disease, an incurable, fatal neurological (brain) disorder."

What does the sentence "Carroll has seen the future" refer to in this excerpt?

A. Some of Carroll's lab mates made a breakthrough in 2006.

B. Carroll knows he will probably start to show symptoms of Huntington's diease around age 49.

C. Carroll took Internet biology courses in his free time so that he could better understand Huntington's disease.

D. Carroll left the service to return to school and earn a bachelor's degree in biology.

7. Read these sentences from the text.

"The first are usually depression, forgetfulness, clumsiness, and mood swings. As time goes on, mental deterioration sets in and uncontrollable jerking movements develop. People eventually lose the abilities to walk, speak, and care for themselves."

What word could replace the phrase "as time goes on" without changing the meaning?

A. Including
B. Although
C. Ultimately
D. Primarily

8. What does the capase 6 enzyme do to the huntingtin protein?

9. In 2006, some of Carroll's lab mates made a breakthrough in their work with a strain of mice engineered to carry the human gene for Huntington's.

Why was their work considered a breakthrough?

Support your answer with evidence from the text.

10. Read this sentence from the text.

"Carroll is looking for a drug that will protect the faulty huntingtin protein from the cutting enzyme."

Why might a drug that will protect the protein from the cutting enzyme be helpful in finding a cure for the disease? Support your answer with evidence from the text.