Name _____

Date _____ Period ____

Too Young To Be Old

Imagine being ten years old, trapped in a body that each day becomes a bit more shriveled, a bit more frail, *old*. You are just tall enough to peer over the top of the kitchen counter, and you weigh less than thirty-five pounds. Already you are bald, and your nose has become crinkled and beaklike.

Most likely, you have only a few more years to live. Yet in spite of this cruel twist of nature, you still have not lost your courage or your childlike curiosity about life. Like Mickey Hayes and Fransie Geringer (Figure 12.1), you play, laugh, celebrate birthdays, and hug your friends.

Of every 8 million humans born, one is destined to grow old far too soon, compared to the normal timetable for our species. Something has gone wrong with one gene on just one of the forty-six chromosomes brought together by chance at conception. From that moment on, the mistake is perpetuated each time cells of the embryo—then of the child—duplicate their chromosomes and divide. The outcome of that rare mistake will be an acceleration of the aging process and a greatly reduced life expectancy. This is the *Hutchinson-Gilford progeria syndrome*, once called the "leprechaun's disease." There is no cure.

Figure 12.1 Two boys, both less than ten years old, who met at Disneyland, California, during a gathering of progeriacs. Progeria is a genetic disorder characterized by accelerated aging and extremely reduced life expectancy.

Usually, symptoms begin to appear before affected individuals are two years old. In some unknown way, the interaction of the altered gene with other genes has absolutely devastating effects on normal cell division, growth, and development. The rate of growth quickly declines to abnormally low levels. Skin becomes thinner and muscles become flaccid. Limb bones that otherwise should lengthen and become stronger start to soften. Most of the time, all hair is lost and the individuals become bald. Progeriacs never reach puberty, the onset of sexual maturation. Most die in their early teens from a stroke or heart attack brought on by an earlier hardening of the arteries, a condition typical of advanced age.

Outwardly, none of the chromosomes of affected individuals appears to be defective when viewed with a microscope. Simple analysis tells us that the condition probably arises through a dominant gene mutation that strikes arbitrarily. The mutated gene is always expressed. It does not occur on a sex chromosome, because the resulting disorder can occur in either boys or girls. There are no documented cases of progeria running in families. As is the case for some other dominant gene mutations, there is a "paternal age effect," in that the father is four or five years older than the mother at the time of conception, on the average.

Name _____

Date _____ Per. _____

Too Young To Be Old

Right There: (It's in the reading)

1. What is the genetic disorder called that accelerates ageing?
2. At what age do symptoms begin to appear?
3. Can the disorder occur in boys and girls?
4. Does the disorder run in families?

Think and Search: (It's there, but you need to read and understand)

5. Explain the symptoms of the disorder. Use complete sentences.

Author and You: (author mentions subject, what do you know about it?)

Why do you think this disorder is called "Leprechaun's disease?" (Complete sentences!)

On your own: (You didn't need to read the article, but maybe it stirred up some ideas) Write at least a paragraph.

How would you feel if you woke up tomorrow and discovered you were 88 years old?