Progeria

Progeria, <<*proh JIHR ee uh,*>> an extremely rare disorder, causes the appearance of advanced aging in children. It occurs once in every 4 million births. The word *progeria* comes from a Greek word meaning *prematurely old.* Hutchinson-Gilford progeria syndrome (HGPS), a common form of the disease, was first described in 1886 by the English physician Sir Jonathan Hutchinson.

Most children with HGPS appear normal at birth, but their growth soon slows. Their primary teeth *erupt* (push through the gums) later and fall out many years later than normal. By 1 or 2 years of age, their hair starts to fall out. By age 3 or 4, they are nearly bald, with almost no eyebrows. Their noses look “pinched,” and they have small jaws. Eventually, they lose most of their body fat. The skin becomes thin-looking, tight in places, and speckled. Veins are easily seen through the skin. The joints become tight, causing the body to take on a stooped appearance. People with HGPS develop high blood pressure, angina, strokes, and heart attacks. They die of heart disease between the ages of 7 and 21.

The disease does not affect a person’s mental development, and many people with HGPS are highly intelligent. Although people who have HGPS continue to grow slowly, few reach a height of 3 feet 6 inches (107 centimeters) or a weight of 40 pounds (18 kilograms).

In 2003, scientists discovered that a genetic mutation causes HGPS. They identified a gene, called LMNA, that controls the production of a protein known as Lamin A. This protein makes up part of the membrane that surrounds the cell nucleus. Scientists think that the damaged protein makes the cells of the body unstable. This instability leads to the process of premature aging.

There is no effective treatment for HGPS. However, medications and nutritional and physical therapy are used to improve the quality of life as researchers search for new treatments.

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