**What is progeria?**

**HGPS (Hutchinson-Gilford Progeria Syndrome) or Progeria** is an extremely rare, fatal genetic condition. The word Progeria comes from the Greek *progeros* meaning 'prematurely old'. The Greek word *pro* means 'before', while the word *geras*means 'old age'.

**Progeria affects children and gives them an appearance of accelerated aging**. The classic type of Progeria (there are different forms) is Hutchinson-Gilford Progeria Syndrome (HGPS). Progeria was first described in an academic journal by Dr. Jonathan Hutchinson in 1886, and Dr. Hastings Gilford in 1897 - both in England.

**How prevalent is Progeria?**

According to [**Hayley's Page**](http://hayleyspage.com/) *"At present there are 53 known cases of Progeria around the world and only 2 in the UK"*. There is a reported incidence of Progeria of approximately 1 in every 4 to 8 million newborns. Both boys and girls run an equal risk of having Progeria.

Progeria appears to affect children of all races equally. Over the last 15 years the following countries have had reported cases - Algeria, Argentina, Australia, Austria, Canada, China, Cuba, England, France, Germany, Israel, Italy, Mexico, the Netherlands, Poland, Puerto Rico, South Africa, South America, South Korea, Switzerland, Turkey, the US, Venezuela, Vietnam and Yugoslavia.

**What are the characteristics of Progeria?**

 
In Progeria patients, the cell nucleus has dramatically aberrant morphology (bottom, middle) rather than the uniform shape typically found in healthy individuals (top, middle)

Children with Progeria are born looking healthy. When they are about 10 to 24 months old, features of accelerated aging start to appear.

**Signs of Progeria**

* Growth failure (Slowed growth, with below-average height and weight)
* Diminished body fat and muscle
* Hair loss (alopecia), including eyelashes and eyebrows
* Skin starts to look aged
* Stiffness in the joints
* Hip dislocation
* Generalized [**atherosclerosis**](http://www.medicalnewstoday.com/articles/247837) (cardio and [**heart disease**](http://www.medicalnewstoday.com/articles/237191))
* [**Stroke**](http://www.medicalnewstoday.com/articles/7624)
* A narrowed face and beaked nose
* Hardening and tightening of skin on trunk and extremities (scleroderma)
* Head disproportionately large for face
* Thin lips
* Visible veins
* Prominent eyes
* Small lower jaw (micrognathia)
* High-pitched voice
* Delayed and abnormal tooth formation
* Insulin resistance
* Irregular heartbeat

Although they may come from varying ethnic backgrounds, children with Progeria have a surprisingly similar appearance. Progeria patients generally die between the ages of 8 and 21 - with the average age being 13.

**How are Progeria and ageing similar?**

Children who suffer from Progeria are genetically susceptible to premature, progressive heart disease. Nearly all Progeria patients die from heart disease. Heart disease is also one of the leading causes of death globally. Children with Progeria commonly experience cardiovascular events, such as hypertension ([**high blood pressure**](http://www.medicalnewstoday.com/articles/159283)), stroke, [**angina**](http://www.medicalnewstoday.com/articles/8886), enlarged heart and [**heart failure**](http://www.medicalnewstoday.com/articles/156849) - conditions linked to aging.

Other health problems that are frequently associated with aging — such as far-sightedness and Alzheimer's disease — do not develop as part of the course of progeria.

Experts say that any research into finding a cure for Progeria would probably have results which would benefit adults with diseases linked to aging.

**What causes Progeria?**

90% of children with Progeria have a mutation on the gene that encodes Lamin A, a protein that holds the nucleus of the cell together. It is believed that the defective Lamin A protein makes the nucleus unstable. This instability seems to lead to the process of premature aging among Progeria patients.

Progeria appears to occur without cause - it is not seen in siblings of affected children. In extremely rare cases more than one child in the same family may have the condition.

**Is Progeria hereditary?**

Experts do not believe that Progera is hereditary. They say it is due to a rare gene change which happens purely by chance. A non-twin sibling runs the same risk of having Progera as any other child from another family. In about 1 in every 100 cases of HGPS the syndrome is passed down to the next generation within the same family.

**Diagnosis of Progeria**

The health care professional will possibly suspect Progeria if the signs and symptoms are there - aging skin, loss of hair, stiffness of joints, etc. This can then be confirmed through a genetic test. The **[Progeria Research Foundation](http://www.progeriaresearch.org/%22%20%5Ct%20%22_blank)** has created a Diagnostic Testing Program.

**Treatment hope**

Farnesyltransferase inhibitors (FTIs), currently used for treating [**cancer**](http://www.medicalnewstoday.com/info/cancer-oncology/), might reverse the nuclear structure abnormalities that are believed to cause Progeria. Studies carried out on mice with Progeria-like signs and symptoms showed that FTIs appeared to offer some improvements. Of the 13 mice treated with FTI, only one died during the 20-week UCLS study. Dr Leslie Gordon, director of the Progeria Research Foundation, said: "This study gives us pieces of information critical to our movement toward clinical trials in children with progeria."

In September 2012, results were released for a [**first clinical drug trial treatment for children with Progeria, using a farnesyltransferase inhibitor**](http://www.medicalnewstoday.com/articles/250725.php). The clinical trial results showed significant improvements in bone structure, weight gain, and the cardiovascular system.

**Coping and support**

Learning your child has progeria can be emotionally devastating. Suddenly you know that your child is facing numerous, difficult challenges and a shortened life span. For you and your family, coping with the disorder involves a major commitment of physical, emotional and financial resources.

In dealing with a disorder such as progeria, support groups can be a valuable part of a wider network of social support that includes health care professionals, family and friends. In a support group, you'll be with people who are facing challenges similar to the one that you are. Talking to group members can help you cope with your own feelings about your child's condition. If a group isn't for you, talking to a therapist or clergy member may be beneficial.

If your child has progeria, he or she is also likely to experience fear and grief as awareness grows that progeria shortens life span. Your child eventually will need your help coping with the concept of death, and may have a number of difficult but important questions about spirituality and religion. Your child may also ask questions about what will happen in your family after he or she dies.

It's critical that you are able to talk openly and honestly with your child, and offer reassurance that's compatible with your belief system. Friends who you meet through support groups also may be able to offer valuable guidance. Your child might also benefit from talking to a therapist or clergy member.

**Sources:**

Written by Christian Nordqvist, Article updated 26 September 2012, **Copyright:** Medical News Today

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