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Premature aging syndromes (progeria)

Premature aging syndromes, also known as progeria, include two very rare inherited conditions, Hutchinson–Gilford syndrome and Werner syndrome. In both conditions, skin changes that indicate premature aging include:

- atrophy (skin thinning and loss of elasticity)
- loss of cutaneous fat
- wrinkling
- greying hair
- loss of hair
- nail dystrophy
- defective pigmentation
- ulceration.

These are changes that occur as the normal body ages (see [ageing skin](#)) but in progeria they occur at an accelerated rate.

What are the differences between progeria types?

Hutchinson–Gilford Syndrome	Werner Syndrome
Occurs in about 1 in 8 million children	Occurs in about 1 in 1 million individuals
Signs of progeria begin to show around 6–12 months when the baby fails to gain weight and skin changes occur	First sign of syndrome is around puberty when the child fails to have a normal growth spurt, or may be delayed until an individual is as old as 30 years
Characteristic features include: <ul style="list-style-type: none"> • Baldness, prominent scalp, veins and eyes, small jaw, delayed tooth formation • Thin limbs with prominent joints, short stature, joint stiffness, hip dislocations • Thickened, tight and shiny skin over joints 	Characteristic features include: <ul style="list-style-type: none"> • Striking difference between person's appearance and his/her real age • Greying hair and/or balding, wrinkling and aging of the face, sunken cheeks, small jaw • Small stature (usually less than 1.6m tall), muscle weakness • Thickened, tight and shiny skin over joints, leading to ulcers • High-pitched voice
Average life expectancy is 13 years, with approximately 75% dying from heart disease	Death usually occurs in patients between 30–50 years, with most dying from heart disease or cancer

What is the cause of progeria?

Both Hutchinson–Gilford syndrome and Werner syndrome are autosomal recessive disorders, which means an individual has inherited a mutated gene from both parents. 1 in 4 offspring would be expected to have the disorder and others may be carriers of the gene.

Werner syndrome appears to be caused by a defect at the WS gene (WRN) locus, which provides instructions to a protein called helicase. Werner syndrome is caused by a helicase defect, which affects the way DNA and RNA are replicated and repaired in the body.

The cause of Hutchinson–Gilford has only been discovered recently. It has been found that there is a mutation in the gene LMNA that produces the protein Lamin A, which is the structural scaffolding that holds the nucleus of a cell together. The cellular instability appears to lead to premature aging.

What are the complications of progeria?

Complications of progeria are related to the diseases that are associated with ageing. In addition to skin changes, patients with Werner syndrome often develop the following conditions:

- Cataracts – rapidly progressing cataracts develop between the ages of 20–40 years in most cases
- Osteoporosis (loss of bone density) – particularly in the legs, caused by disturbances in the parathyroid glands
- Diabetes mellitus – this occurs in at least 30% of patients, and there is abnormal glucose tolerance in many others
- Cardiovascular disease – particularly arteriosclerotic disease, which may lead to fatal heart attack
- Pituitary gland underactivity
- Hypogonadism or gonadism (absent or underactive ovaries or testes) and premature menopause
- Soft tissue calcification
- Cancer – the incidence of malignancy is high, especially fibrosarcomas which occur in 10% of patients. Other cancers include carcinomas of the thyroid and other organs, blood disorders, meningiomas and [skin cancers](#) including [squamous cell carcinoma](#) arising in ankle and heel ulcers.

What is the treatment for progeria?

There is no specific treatment for progeria. Management of the syndromes is through symptomatic treatment of related diseases. Genetic counselling is very important.

Related information

References:

- Book: Textbook of Dermatology. Ed Rook A, Wilkinson DS, Ebling FJB, Champion RH, Burton JL. Fourth edition. Blackwell Scientific Publications.

On DermNet NZ:

- [Cutaneous markers of internal malignancy](#)

Other websites:

- emedicine dermatology, the online textbook:
 - [Progeria \(Werner Syndrome\)](#)
 - [Hutchinson–Gilford Progeria](#)

Books about skin diseases:

See the [DermNet NZ bookstore](#)

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DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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