[Template:Infobox medical condition](/wiki/Template:Infobox_medical_condition" \o "Template:Infobox medical condition)

**Progeria** ([Template:IPAc-en](/wiki/Template:IPAc-en)[[1]](#cite_note-1)[[2]](#cite_note-2)) (**Hutchinson–Gilford progeria syndrome**,[[3]](#cite_note-3)[[4]](#cite_note-4) **HGPS**, **progeria syndrome**[[4]](#cite_note-4)) is an extremely rare [genetic disorder](/wiki/Genetic_disorder) in which symptoms resembling aspects of [aging](/wiki/Senescence) are manifested at a very early age.[[5]](#cite_note-5) Progeria is one of several [progeroid syndromes](/wiki/Progeroid_syndromes).[[6]](#cite_note-6) The word *progeria* comes from the Greek words "pro" ([πρό](/wiki/Wikt:πρό)), meaning "before" or "premature", and "gēras" ([γῆρας](/wiki/Wikt:γῆρας)), meaning "old age".[[7]](#cite_note-7)[[12]](#cite_note-12) Progeria was first described in 1886 by [Jonathan Hutchinson](/wiki/Jonathan_Hutchinson).[[13]](#cite_note-13) It was also described independently in 1897 by [Hastings Gilford](/wiki/Hastings_Gilford).[[14]](#cite_note-14) The condition was later named Hutchinson–Gilford progeria syndrome.

## Contents

* 1 Signs and symptoms[[edit](/index.php?title=(none)&action=edit&section=1)]
* 2 Cause[[edit](/index.php?title=(none)&action=edit&section=2)]
* 3 Diagnosis[[edit](/index.php?title=(none)&action=edit&section=3)]
* 4 Treatment[[edit](/index.php?title=(none)&action=edit&section=4)]
* 5 Prognosis[[edit](/index.php?title=(none)&action=edit&section=5)]
* 6 Epidemiology[[edit](/index.php?title=(none)&action=edit&section=6)]
* 7 Research[[edit](/index.php?title=(none)&action=edit&section=7)]
  + 7.1 Lamin A[[edit](/index.php?title=(none)&action=edit&section=8)]
  + 7.2 Mouse model[[edit](/index.php?title=(none)&action=edit&section=9)]
  + 7.3 DNA repair[[edit](/index.php?title=(none)&action=edit&section=10)]
* 8 Popular culture[[edit](/index.php?title=(none)&action=edit&section=11)]

## Signs and symptoms[[edit](/index.php?title=(none)&action=edit&section=1)]

Children with progeria usually develop the first symptoms during their first few months of life. The earliest symptoms may include a [failure to thrive](/wiki/Failure_to_thrive) and a localized [scleroderma](/wiki/Scleroderma)-like skin condition. As a child ages past infancy, additional conditions become apparent usually around 18–24 months. Limited growth, full-body [alopecia](/wiki/Alopecia) (hair loss), and a distinctive appearance (a small face with a shallow recessed jaw, and a pinched nose) are all characteristics of progeria. Signs and symptoms of this progressive disease tend to become more marked as the child ages. Later, the condition causes wrinkled skin, [atherosclerosis](/wiki/Atherosclerosis), kidney failure, loss of eyesight, and [cardiovascular](/wiki/Cardiovascular) problems. Scleroderma, a hardening and tightening of the skin on trunk and extremities of the body, is prevalent. People diagnosed with this disorder usually have small, fragile bodies, like those of elderly people. The face is usually wrinkled, with a larger head in relation to the body, a narrow face and a beak nose. Prominent scalp veins are noticeable (made more obvious by alopecia), as well as prominent eyes. Musculoskeletal degeneration causes loss of body fat and muscle, stiff joints, hip dislocations, and other symptoms generally absent in the non-elderly population. Individuals usually retain typical mental and motor development.

## Cause[[edit](/index.php?title=(none)&action=edit&section=2)]

|  |  |
| --- | --- |
| **Steps in normal cell** | **Steps in cell with progeria** |
| The gene [LMNA](/wiki/LMNA) encodes a protein called prelamin A. | |
| Prelamin A has a [farnesyl group](/wiki/Farnesyl_group) attached to its end. | |
| Farnesyl group is *removed* from prelamin A. | Farnesyl group remains *attached* to prelamin A. |
| Normal form is called [lamin A](/wiki/LMNA). | Abnormal form of prelamin A is called [progerin](/wiki/Progerin). |
| Lamin A is not anchored to the nuclear rim. | Progerin is anchored to the nuclear rim. |
| Normal state of the nucleus. | Abnormally shaped nucleus. |

In normal conditions, the [*LMNA*](/wiki/LMNA) gene codes for a structural protein called prelamin A which undergoes a series of processing steps before becoming its final form, called lamin A.<ref name=ghr>[LMNA](http://ghr.nlm.nih.gov/gene/LMNA) [Template:Color](/wiki/Template:Color) [Genes](http://ghr.nlm.nih.gov/BrowseGenes) [Template:Color](/wiki/Template:Color) [Genetics Home Reference](http://ghr.nlm.nih.gov/)</ref> In one of these steps, after prelamin A is [made](/wiki/Translation_(biology)) in the [cytoplasm](/wiki/Cytoplasm), an [enzyme](/wiki/Enzyme) called [farnesyl transferase](/wiki/Farnesyl_transferase) [attaches a farnesyl](/wiki/Prenylation) [functional group](/wiki/Moiety_(chemistry)) to its [carboxyl-terminus](/wiki/Carboxyl-terminus). The [farnesylated](/wiki/Farnesylation) prelamin A is then [transported](/wiki/Protein_transport) through a [nuclear pore](/wiki/Nuclear_pore) to the [interior of the nucleus](/wiki/Nucleoplasm). The farnesyl group allows prelamin A to attach temporarily to the [nuclear rim](/wiki/Nuclear_rim).[Template:Citation needed](/wiki/Template:Citation_needed) Once the protein is attached, it is [cleaved](/wiki/Proteolysis) by a [protease](/wiki/Protease), thereby removing the farnesyl group along with a few adjacent amino acids. Failure to remove this farnesyl group permanently affixes the protein to the nuclear rim. After cleavage by the protease, prelamin A is referred to as [lamin A](/wiki/Lamin_A). Lamin A, along with lamin B and lamin C, makes up the [nuclear lamina](/wiki/Nuclear_lamina), which provides structural support to the nucleus.

Before the late 20th century, research on progeria yielded very little information about the syndrome. In 2003, the cause of progeria was discovered to be a [point mutation](/wiki/Point_mutation) in position 1824 of the *LMNA* gene, in which cytosine is replaced with thymine.[[15]](#cite_note-15) This mutation creates a 5' [cryptic splice site](/wiki/Cryptic_splice_site) within [exon](/wiki/Exon) 11, resulting in an abnormally short mature mRNA transcript. This mRNA strand, when [translated](/wiki/Translation_(biology)), yields an abnormal variant of the prelamin A protein whose farnesyl group cannot be removed. Because its farnesyl group cannot be removed, this abnormal protein, referred to as [progerin](/wiki/Progerin), is permanently affixed to the nuclear rim, and therefore does not become part of the nuclear lamina. Without lamin A, the nuclear lamina is unable to provide the [nuclear envelope](/wiki/Nuclear_envelope) with adequate structural support, causing it to take on an abnormal shape.[[16]](#cite_note-16) Since the support that the nuclear lamina normally provides is necessary for the organizing of [chromatin](/wiki/Chromatin) during [mitosis](/wiki/Mitosis), weakening of the nuclear lamina limits the ability of the cell to divide.[[17]](#cite_note-17) To date over 1,400 [SNPs](/wiki/Single_Nucleotide_Polymorphism) of LMNA gene are known.[[18]](#cite_note-18) They can manifest in changes on mRNA, splicing or protein (e.g. Arg471Cys,[[19]](#cite_note-19) Arg482Gln,[[20]](#cite_note-20) Arg527Leu,[[21]](#cite_note-21) Arg527Cys,[[22]](#cite_note-22) Ala529Val[[23]](#cite_note-23)) level.

Progerin may also play a role in normal human [aging](/wiki/Aging), since its production is activated in typical [senescent](/wiki/Senescence) cells.[[17]](#cite_note-17) Unlike "[accelerated aging diseases](/wiki/DNA_repair-deficiency_disorder)" (such as [Werner syndrome](/wiki/Werner_syndrome), [Cockayne syndrome](/wiki/Cockayne_syndrome) or [xeroderma pigmentosum](/wiki/Xeroderma_pigmentosum)), progeria may not be directly caused by defective [DNA repair](/wiki/DNA_repair). Because these diseases cause changes in different aspects of aging, but never in every aspect, they are often called "segmental progerias."[[24]](#cite_note-24)

## Diagnosis[[edit](/index.php?title=(none)&action=edit&section=3)]

Diagnosis is suspected according to signs and symptoms, such as skin changes, abnormal growth, and loss of hair. A genetic test for LMNA mutations can confirm the diagnosis of progeria.[[25]](#cite_note-25)[[26]](#cite_note-26)

## Treatment[[edit](/index.php?title=(none)&action=edit&section=4)]

No treatment has proven effective. Most treatment focuses on reducing complications (such as [cardiovascular disease](/wiki/Cardiovascular_disease)) with [coronary artery bypass surgery](/wiki/Coronary_artery_bypass_surgery) or low-dose [aspirin](/wiki/Aspirin).[[27]](#cite_note-27) Children may also benefit from a high-energy diet.

[Growth hormone treatment](/wiki/Growth_hormone_treatment) has been attempted.[[28]](#cite_note-28) The use of [Morpholinos](/wiki/Morpholino) has also been attempted in order to reduce progerin production. Antisense Morpholino oligonucleotides specifically directed against the mutated exon 11–exon 12 junction in the mutated pre-mRNAs were used.[[29]](#cite_note-29) [thumb|Potential therapeutic targets for the inhibition of progerin farnesylation](/wiki/File:Medicationsthatinhibitfarnesylation.jpeg) A type of anticancer drug, the [farnesyltransferase inhibitors](/wiki/Farnesyltransferase_inhibitor) (FTIs), has been proposed, but their use has been mostly limited to [animal models](/wiki/Animal_model).[[30]](#cite_note-30) A Phase II clinical trial using the FTI [lonafarnib](/wiki/Lonafarnib) began in May 2007.[[31]](#cite_note-31) In studies on the cells another anti-cancer drug, [rapamycin](/wiki/Rapamycin), caused removal of [progerin](/wiki/Progerin) from the nuclear membrane through [autophagy](/wiki/Autophagy).[[16]](#cite_note-16)[[32]](#cite_note-32) It has been proved that [pravastatin](/wiki/Pravastatin) and [zoledronate](/wiki/Zoledronate) are effective drugs when it comes to the blocking of farnesyl group production.

Farnesyltransferase inhibitors (FTIs) are drugs that inhibit the activity of an enzyme needed in order to make a link between progerin proteins and farnesyl groups. This link generates the permanent attachment of the progerin to the nuclear rim. In progeria, cellular damage can be appreciated because that attachment takes place and the nucleus is not in a normal state. Lonafarnib is an FTI, which means it can avoid this link, so progerin can not remain attached to the nucleus rim and it now has a more normal state.

The delivery of [Lonafarnib](/wiki/Lonafarnib) is not approved by the US [Food and Drug Administration](/wiki/Food_and_Drug_Administration) (FDA). Therefore, it can only be used in certain clinical trials. Until the treatment of FTIs is implemented in progeria children we will not know its effects—which are positive in mice.[[33]](#cite_note-33) [Pravastatin](/wiki/Pravastatin), traded as Pravachol or Selektine, is included in the family of statins. As well as [zoledronate](/wiki/Zoledronate) (also known as Zometa and Reclast, which is a bisphosphonate), its utility in HGPS is the prevention of farnesyl group formation, which progerin needs to provoke the disease. Some animal trials have been realized using FTIs or a combination of [pravastatin](/wiki/Pravastatin) and [zoledronate](/wiki/Zoledronate) so as to observe whether they are capable of reversing abnormal nuclei.

The results, obtained by blinded electron microscopic analysis and immunofluorescence microscopy, showed that nucleus abnormalities could be reversed in transgenic mice expressing progerin. The reversion was also observed in vivo—cultured cells from human subjects with progeria—due to the action of the pharmacs, which block protein prenylation (transfer of a farnesyl polypeptide to C-terminal cysteine). The authors of that trial add, when it comes to the results, that: "They further suggest that skin biopsy may be useful to determine if protein farnesylation inhibitors are exerting effects in subjects with HGPS in clinical trials".[[34]](#cite_note-34) Unlike FTIs, [pravastatin](/wiki/Pravastatin) and [zoledronate](/wiki/Zoledronate) were approved by the U.S. FDA (in 2006 and 2001 respectively), although they are not sold as a treatment for progeria. [Pravastatin](/wiki/Pravastatin) is used to decrease cholesterol levels and [zoledronate](/wiki/Zoledronate) to prevent [hypercalcaemia](/wiki/Hypercalcaemia).

[Rapamycin](/wiki/Rapamycin), also known as [Sirolimus](/wiki/Sirolimus), is a [macrolide](/wiki/Macrolide). There are recent studies concerning rapamycin which conclude that it can minimize the phenotypic effects of progeria fibroblasts. Other observed consequences of its use are: abolishment of nuclear blebbing, degradation of progerin in affected cells and reduction of insoluble progerin aggregates formation. All these results do not come from any clinical trial, although it is believed that the treatment might benefit HGPS patients.[[16]](#cite_note-16) A 2012 clinical trial found that the cancer drug [Lonafarnib](/wiki/Lonafarnib) can improve weight gain and other symptoms of progeria.[[35]](#cite_note-35)

## Prognosis[[edit](/index.php?title=(none)&action=edit&section=5)]

As there is no known cure, few people with progeria exceed 13 years of age.[[36]](#cite_note-36) At least 90% of patients die from complications of [atherosclerosis](/wiki/Atherosclerosis), such as heart attack or stroke.[[37]](#cite_note-37) Mental development is not adversely affected; in fact, intelligence tends to be average to above average.[[38]](#cite_note-38) With respect to the features of aging that progeria appears to manifest, the development of symptoms is comparable to aging at a rate eight to ten times faster than normal. With respect to features of aging that progeria does not exhibit, patients show no [neurodegeneration](/wiki/Neurodegeneration) or [cancer](/wiki/Cancer) predisposition. They also do not develop the so-called "wear and tear" conditions commonly associated with aging, such as [cataracts](/wiki/Cataracts) (caused by UV exposure) and [osteoarthritis](/wiki/Osteoarthritis) (caused by mechanical wear).[[25]](#cite_note-25) Although there may not be any successful treatments for progeria itself, there are treatments for the problems it causes, such as arthritic, respiratory, and cardiovascular problems. Sufferers of progeria have normal reproductive development and there are known cases of women with progeria who had delivered healthy offspring.[[39]](#cite_note-39)

## Epidemiology[[edit](/index.php?title=(none)&action=edit&section=6)]

A study from the Netherlands has shown an incidence of 1 in 4 million births.[[40]](#cite_note-40) Currently, there are 100 known cases in the world. Approximately 140 cases have been reported in medical history.[[41]](#cite_note-41) However, the [Progeria Research Foundation](/wiki/Progeria_Research_Foundation) believes there may be as many as 150 undiagnosed cases worldwide.

Classical Hutchinson–Gilford progeria syndrome is usually caused by a sporadic mutation taking place during the early stages of embryo development. It is almost never passed on from affected parent to child, as affected children rarely live long enough to have children themselves.

There have been only two cases in which a healthy person was known to carry the LMNA mutation that causes progeria. These carriers were identified because they passed it on to their children.<ref name=Kork08/> One family from India has five children with progeria, though not the classical HGPS type.[[42]](#cite_note-42) This family was the subject of a 2005 [Bodyshock](/wiki/Bodyshock) documentary entitled [*The 80 Year Old Children*](/wiki/The_80_Year_Old_Children). The Vandeweert family of Belgium has two children, Michiel and Amber, with classic HGPS.[[43]](#cite_note-43)

## Research[[edit](/index.php?title=(none)&action=edit&section=7)]

Several discoveries have been made that have led to greater understandings and perhaps eventual treatment for this disease.[[44]](#cite_note-44)[[45]](#cite_note-45) A 2003 report in *Nature*[[46]](#cite_note-46) said that progeria may be a [de novo](/wiki/De_novo_mutation) [dominant trait](/wiki/Dominant_trait). It develops during [cell division](/wiki/Cell_division) in a newly conceived zygote or in the [gametes](/wiki/Gamete) of one of the parents. It is caused by [mutations](/wiki/Mutation) in the LMNA ([lamin A](/wiki/Lamin_A) [protein](/wiki/Protein)) [gene](/wiki/Gene) on [chromosome 1](/wiki/Chromosome_1); the mutated form of lamin A is commonly known as [progerin](/wiki/Progerin). One of the authors, Leslie Gordon, was a physician who did not know anything about progeria until her own son, [Sam](/wiki/Sam_Berns), was diagnosed at 22 months. Gordon and her husband, pediatrician Scott Berns, founded the [Progeria Research Foundation](/wiki/Progeria_Research_Foundation).[[47]](#cite_note-47)

### Lamin A[[edit](/index.php?title=(none)&action=edit&section=8)]

[Lamin A](/wiki/Lamin_A) is a major component of a protein [scaffold](/wiki/Scaffold) on the inner edge of the [nucleus](/wiki/Cell_nucleus) called the [nuclear lamina](/wiki/Nuclear_lamina) that helps organize nuclear processes such as [RNA](/wiki/RNA) and [DNA](/wiki/DNA) synthesis.

Prelamin A contains a [CAAX box](/wiki/CAAX_box) at the [C-terminus](/wiki/C-terminus) of the protein (where C is a [cysteine](/wiki/Cysteine) and A is any [aliphatic](/wiki/Aliphatic) [amino acids](/wiki/List_of_standard_amino_acids)). This ensures that the cysteine is [farnesylated](/wiki/Prenylation) and allows prelamin A to bind [membranes](/wiki/Cell_membrane), specifically the nuclear membrane. After prelamin A has been localized to the cell nuclear membrane, the C-terminal amino acids, including the farnesylated cysteine, are cleaved off by a specific [protease](/wiki/Protease). The resulting protein, now lamin A, is no longer membrane-bound, and carries out functions inside the nucleus.

In HGPS, the recognition site that the enzyme requires for cleavage of prelamin A to lamin A is mutated. Lamin A cannot be produced, and prelamin A builds up on the nuclear membrane, causing a characteristic nuclear [blebbing](/wiki/Blebbing).[[48]](#cite_note-48) This results in the symptoms of progeria, although the relationship between the misshapen nucleus and the symptoms is not known.

A study that compared HGPS patient cells with the skin cells from young and elderly normal human subjects found similar defects in the HGPS and elderly cells, including [down-regulation](/wiki/Down-regulation) of certain nuclear proteins, increased [DNA](/wiki/DNA) damage, and [demethylation](/wiki/Demethylation) of [histone](/wiki/Histone), leading to reduced [heterochromatin](/wiki/Heterochromatin).[[49]](#cite_note-49) [Nematodes](/wiki/Nematode) over their lifespan show progressive lamin changes comparable to HGPS in all cells but [neurons](/wiki/Neuron) and [gametes](/wiki/Gamete).[[50]](#cite_note-50) These studies suggest that lamin A defects are associated with normal [aging](/wiki/Senescence). [Template:Clear](/wiki/Template:Clear)

### Mouse model[[edit](/index.php?title=(none)&action=edit&section=9)]

[thumb|left|250px|Confocal microscopy photographs of the descending aortas of two 15-month-old progeria mice, one untreated (left) and the other treated with the FTI drug tipifarnib (right)](/wiki/File:Progeria20132-300.jpg) [thumb|200px|Untreated cells from children with the genetic disease progeria (left) compared to similar cells treated with FTIs](/wiki/File:Progeria37-72.jpg)

A mouse [model](/wiki/Animal_model) of progeria exists, though in the mouse, the [LMNA](/wiki/LMNA) prelamin A is not mutated. Instead, [ZMPSTE24](/wiki/ZMPSTE24), the specific protease that is required to remove the C-terminus of prelamin A, is missing. Both cases result in the buildup of farnesylated prelamin A on the nuclear membrane and in the characteristic nuclear LMNA blebbing. Fong et al. use a [farnesyl transferase inhibitor](/wiki/Farnesyl_transferase_inhibitor) (FTI) in this mouse model to inhibit protein farnesylation of prelamin A. Treated mice had greater grip strength and lower likelihood of [rib fracture](/wiki/Rib_fracture) and may live longer than untreated mice.[[51]](#cite_note-51) This method does not directly "cure" the underlying cause of progeria. This method prevents prelamin A from going to the nucleus in the first place so that no prelamin A can build up on the nuclear membrane, but equally, there is no production of normal lamin A in the nucleus. Lamin A does not appear to be necessary for life; mice in which the *Lmna* gene is knocked out show no embryological symptoms (they develop an [Emery–Dreifuss muscular dystrophy](/wiki/Emery–Dreifuss_muscular_dystrophy)-like condition postnatally).[[52]](#cite_note-52) This implies that it is the buildup of prelamin A in the wrong place, rather than the loss of the normal function of lamin A, that causes the disease.

It was hypothesized that part of the reason that treatment with an FTI such as [alendronate](/wiki/Alendronate) is inefficient is due to [prenylation](/wiki/Prenylation) by [geranylgeranyltransferase](/wiki/Geranylgeranyltransferase). Since [statins](/wiki/Statins) inhibit geranylgeranyltransferase, the combination of an FTI and statins was tried, and markedly improved "the aging-like phenotypes of mice deficient in the metalloproteinase Zmpste24, including growth retardation, loss of weight, lipodystrophy, hair loss, and bone defects".[[53]](#cite_note-53)[Template:Clear](/wiki/Template:Clear)

### DNA repair[[edit](/index.php?title=(none)&action=edit&section=10)]

Repair of DNA double-strand breaks can occur by either of two processes, [non-homologous end joining](/wiki/Non-homologous_end_joining) (NHEJ) or [homologous recombination](/wiki/Homologous_recombination) (HR). A-type [lamins](/wiki/Lamin) promote genetic stability by maintaining levels of proteins that have key roles in NHEJ and HR.[[54]](#cite_note-54) Mouse cells deficient for maturation of prelamin A show increased DNA damage and [chromosome aberrations](/wiki/Chromosome_abnormality) and have increased sensitivity to DNA damaging agents.[[55]](#cite_note-55) In progeria, the inability to adequately repair DNA damages due to defective A-type lamin may cause aspects of premature aging[[56]](#cite_note-56) (also see [DNA damage theory of aging](/wiki/DNA_damage_theory_of_aging)).

## Popular culture[[edit](/index.php?title=(none)&action=edit&section=11)]

Perhaps one of the earliest influences of progeria on popular culture occurred in the 1922 short story [*The Curious Case of Benjamin Button*](/wiki/The_Curious_Case_of_Benjamin_Button_(short_story)) by [F. Scott Fitzgerald](/wiki/F._Scott_Fitzgerald) (and later released as a [feature film](/wiki/The_Curious_Case_of_Benjamin_Button_(film)) in 2008). The main character, Benjamin Button, is born as a seventy-year-old man and ages backwards; it has been suggested that this was inspired by progeria.[[57]](#cite_note-57) [Charles Dickens](/wiki/Charles_Dickens) may have described a case of progeria in the Smallweed family of [*Bleak House*](/wiki/Bleak_House), specifically in the grandfather and his grandchildren, Judy and twin brother Bart.[[58]](#cite_note-58) A Bollywood movie, [*Paa*](/wiki/Paa_(film)), was made about the condition; in it, the lead ([Amitabh Bachchan](/wiki/Amitabh_Bachchan)) played a 13-year-old child affected by progeria.

In the 1983 film [*The Hunger*](/wiki/The_Hunger_(1983_film)), progeria was the focus of study by [Susan Sarandon's](/wiki/Susan_Sarandon) character, Dr. Sarah Roberts.

The 1984 film [*The Three Wishes of Billy Grier*](/wiki/The_Three_Wishes_of_Billy_Grier) stars [Ralph Macchio](/wiki/Ralph_Macchio) as a teenager who tries to fulfill his wishes before he dies from the disease.

The 1996 movie [*Jack*](/wiki/Jack_(1996_film)) deals with the eponymous character ([Robin Williams](/wiki/Robin_Williams)) who has a genetic disorder similar to progeria and the difficulties he faces fitting into society.

The 2006 movie [*Renaissance*](/wiki/Renaissance_(film)) deals with progeria.

["Young at Heart"](/wiki/Young_at_Heart_(The_X-Files)), the sixteenth episode of the first season of the television show [*The X-Files*](/wiki/The_X-Files), features a violent criminal who has seemingly grown younger due to treatment by a corrupt doctor, who had developed his technique by experimenting on progeria sufferers.

In Tad Williams' novel series [*Otherland*](/wiki/Otherland), one of the main characters suffers from progeria.

Harold Kushner's 1978 book [*When Bad Things Happen to Good People*](/wiki/When_Bad_Things_Happen_to_Good_People), which explores God and the problem of evil, was written in response to his 14-year-old son's death due to progeria.

South African artist/hip hop artist [Leon Botha](/wiki/Leon_Botha) was one of the oldest known progeria sufferers, surviving to the age of 26 before his death in June 2011.

Meg Casey, a Milford, CT artist and spokesperson for the handicapped, was born October 1, 1955 and died May 26, 1985. She survived for 29 years with progeria.[[59]](#cite_note-59)