

Progeria, or Hutchinson-Gilford progeria syndrome (HGPS), is a rare, fatal, genetic condition of childhood with striking features resembling premature aging. Children with progeria usually have a normal appearance in early infancy. At approximately nine to 24 months of age, affected children begin to experience profound growth delays, resulting in short stature and low weight. They also develop a distinctive facial appearance characterized by a disproportionately small face in comparison to the head; an underdeveloped jaw (micrognathia); malformation and crowding of the teeth; abnormally prominent eyes; a small nose; prominent eyes and a subtle blueness around the mouth. In addition, by the second year of life, the scalp hair, eyebrows, and eyelashes are lost (alopecia), and the scalp hair may be replaced by small, downy, white or blond hairs. Additional characteristic features include generalized atherosclerosis, cardiovascular disease and stroke, hip dislocations, unusually prominent veins of the scalp, loss of the layer of fat beneath the skin (subcutaneous adipose tissue), defects of the nails, joint stiffness, skeletal defects, and/or other abnormalities. According to reports in the medical literature, individuals with HGPS develop premature, widespread thickening and loss of elasticity of artery walls (arteriosclerosis), which result in life-threatening complications during childhood, adolescence, or early adulthood. Children with progeria die of heart disease (atherosclerosis) at an average age of 13 years, with a range of about eight to 21 years. As with any person suffering from heart disease, the common events as heart disease advances for children with progeria can include high blood pressure, strokes, angina (chest pain due to poor blood flow to the heart itself), enlarged heart, and heart failure, all conditions associated with aging. HGPS is a rare disorder that appears to affect males and females equally, and all races equally. The disorder was originally described in the medical literature in 1886 (J. Hutchinson) and 1897 (H. Gilford). As of January 2014, approximately 200 cases have been reported. Estimates indicate that the prevalence of HGPS is approximately one in eighteen million, thus at any given time, there are approximately 350-400 children living with progeria worldwide. Two sets of affected identical twins have been reported in the literature.