

Cockayne Syndrome (CS) is a rare form of dwarfism. It is an inherited disorder whose diagnosis depends on the presence of three signs (1) growth retardation, i.e. short stature, (2) abnormal sensitivity to light (photosensitivity), and (3) prematurely aged appearance (progeria). In the classical form of Cockayne Syndrome (CS type I) the symptoms are progressive and typically become apparent after the age of one year. An early onset or congenital form of Cockayne Syndrome (CS type II) is apparent at birth (congenital). There is a third form, known as Cockayne Syndrome Type III (CS type III), that presents later in the child's development and is generally a milder form of the disease. A fourth form; now recognized as Xeroderma pigmentosa-Cockayne syndrome (XP-CS), combines features of both of these disorders. Cockayne Syndrome is very rare and affects males and females in equal numbers. There are no indications of ethnic or racial partiality. The incidence of CS is less than 1 case per 250,000 live births in the U.S. As of 1992, about 140 cases of CS had been reported in the literature.