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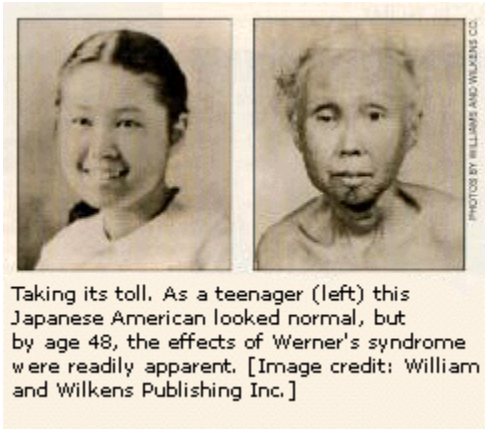
Werner syndrome



Taking its toll. As a teenager (left) this Japanese American looked normal, but by age 48, the effects of Werner's syndrome were readily apparent. [Image credit: William and Wilkens Publishing Inc.]

Werner syndrome is a premature aging disease that begins in adolescence or early adulthood and results in the appearance of old age by 30-40 years of age. Its physical characteristics may include short stature (common from childhood on) and other features usually developing during adulthood: wrinkled skin, baldness, cataracts, muscular atrophy and a tendency to diabetes mellitus, among others.

The disorder is inherited and transmitted as an autosomal recessive trait. Cells from WS patients have a shorter lifespan in culture than do normal cells. The gene for Werner disease (WRN) was mapped to chromosome 8 and cloned: by comparing its sequence to existing sequences in GenBank, it is a predicted helicase belonging to the RecQ family. However, it has yet to be shown to have real helicase activity (as a DNA unwinder important for DNA replication). The molecular role of WRN in Werner syndrome therefore remains to be proven, as does any role it might have in the aging process in general.

A yeast protein similar to the human WRN protein, called SGS1, has been found. Mutations in SGS1 cause yeast to have a shorter lifespan than yeast cells without the mutation, and shown other signs typical of aging in yeast, such as an enlarged and fragmented nucleolus. Using yeast as a model for human aging in general, may give insight into the mechanisms of Werner syndrome and related diseases.

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