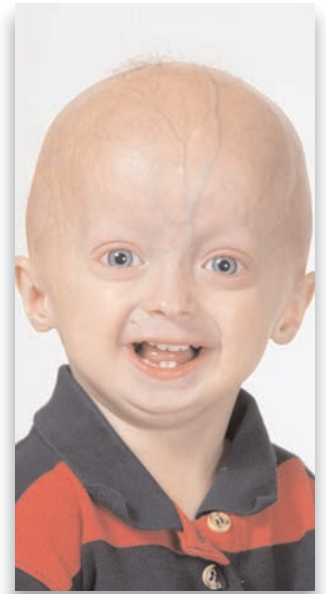


18. Progeria and Aging:

What Progeria and aging have in common and how they are different



Progeria is called a “segmental” premature aging syndrome. That is because it does not mimic aging completely. For example, children with Progeria do not experience Alzheimer’s disease, cataracts, or cancers typical of aging. Conversely, aging in the general population does not bring about some of the bone changes and balding patterns seen in Progeria. It is very important to determine where aging and Progeria overlap at the biological level, so that we can learn and help everyone as much as possible.

Understanding Progeria promises exciting new avenues for understanding the natural aging process.

What Progeria and aging have in common and how they are different

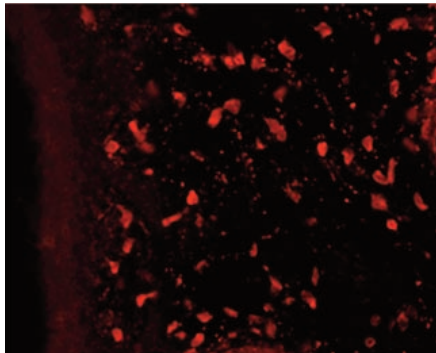
The discovery that Progeria is caused by a newly discovered protein called progerin raised entirely new questions: Is progerin produced by all of us? Does progerin have a role in aging and heart disease? Perhaps our most exciting new clue to the aging process is the discovery that the progerin protein is present at increasing concentrations in both Progeria and normal cells as they age. In addition, progerin is found in skin biopsies of older donors (see figure on next page), while young donors have less or no detectable progerin. The newly discovered relationship between Progeria and progerin has opened the doors of scientific exploration into how this molecule may play a role in heart disease and aging in the general population.

We all make a little bit of progerin, though much less than children with Progeria.

Children with Progeria are genetically predisposed to premature, progressive heart disease. Death occurs almost exclusively due to widespread heart disease, the number one cause of death globally¹.

As with any person suffering from heart disease, the common events for Progeria children are strokes, high blood pressure, angina, enlarged heart, and heart failure, all conditions associated with aging. Thus there is clearly a tremendous need for research in Progeria. Finding a cure for Progeria will not only help these children, but may provide keys for treating millions of adults with heart disease and stroke associated with the natural aging process.

Because the aging process is accelerated in children with Progeria, they offer researchers a rare opportunity to observe in just a few years what would otherwise require decades of longitudinal studies.



Skin biopsy showing progerin in a 93 year-old person without Progeria. The red dots are cells containing progerin. (Photograph courtesy of K. Djabali)

¹World Health Organization; www.who.int/mediacentre/factsheets/fs317/en/index.html