Published in final edited form as:

N Engl J Med. 2008 May 29; 358(22): 2409–2411. doi:10.1056/NEJMc086092.

Hutchinson–Gilford Progeria Syndrome

William B. Ershler, M.D., **Luigi Ferrucci, M.D., Ph.D.**, and **Dan L. Longo, M.D.**

National Institute on Aging, Baltimore, MD 21225

To the Editor

In an otherwise elegant clinical description of the Hutchinson–Gilford progeria syndrome by Merideth and colleagues (Feb. 7 issue),[1] we take exception to the authors' suggestion that their findings have overarching significance with respect to an understanding of normal aging. Although some of the changes that characterize the Hutchinson–Gilford progeria syndrome look like aging, other characteristic features of the syndrome (e.g., elevated platelet counts, prolonged prothrombin times, and functional oral deficits) do not. Moreover, certain prominent features of human aging — for example, the activation of inflammatory pathways — are not typical of the Hutchinson–Gilford progeria syndrome.

On the basis of studies in mice, it is likely that every organ undergoes distinct molecular changes with age.[2–4] Authentic genetic models of accelerated aging should not only give rise to phenotypes that mimic aging but also produce changes in gene expression that are typical of normal aging. This is not the case for the Hutchinson–Gilford progeria syndrome or other progeria syndromes. It is time to relinquish the myth that the Hutchinson–Gilford progeria syndrome and related "progeria" syndromes hold the key to an understanding of aging. This is a convenient and hopeful conclusion, but it is inconsistent with what is known about aging.

References

1. Merideth MA, Gordon LB, Clauss S, et al. Phenotype and course of Hutchinson–Gilford progeria syndrome. N Engl J Med. 2008; 358:592–604. [PubMed: 18256394]
2. Lustig A, Weeraratna AT, Wood WW III, et al. Transcriptome analysis of age-, gender-, and diet-associated changes in murine thymus. Cell Immunol. 2007; 245:42–61. [PubMed: 17499630]
3. Xu X, Zhan M, Duan W, et al. Gene expression atlas of the mouse central nervous system: impact and interactions of age, energy intake and gender. Genome Biol. 2007; 8:R234. [PubMed: 17988385]
4. Zahn JM, Poosala S, Owen AB, et al. AGEMAP: a gene expression database for aging in mice. PLoS Genet. 2007; 3(11):e201. [PubMed: 18081424]