Protein May Increase Lifespan In Premature Aging, Heart Disease

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http://www.asianscientist.com/in-the-lab/sun1-lifespan-progeria-emery-dreifuss-muscular-dystrophy-2012/

AsianScientist (May 2, 2012) - Scientists have discovered that they can dramatically increase the life span of mice with premature aging disease and heart disease by reducing levels of a protein called SUN1.

This research, published recently in the journal *Cell*, was a collaboration among the A*STAR Institute of Medical Biology (IMB) in Singapore, the National Institute of Allergy and Infectious Diseases (NIAID) in the United States, and the Institute of Cellular and System Medicine in Taiwan.

Children with progeria suffer symptoms of premature aging and mostly die in their early teens from either heart attack or stroke. Individuals with a specific type of heart disease, Emery-Dreifuss muscular dystrophy (AD-EDMD), suffer from muscle wasting and cardiomyopathy, which eventually leads to heart failure.

Both diseases are caused by mutations in Lamin A, a protein in the membrane surrounding a cell's nucleus which provides mechanical support to the nucleus. SUN1 is a protein also found in the inner nuclear membrane, but there have been no previous studies to show how SUN1 interacts with the Lamin proteins.

To investigate if SUN1 had any involvement in diseases caused by mutations in Lamin A, the scientists inactivated SUN1 in mouse models developed for progeria and AD-EDMD. Instead of the short life spans commonly observed in these mouse models, the mice lived twice (progeria) and thrice (AD-EDMD) as long compared to when SUN1 levels were high.

> "We actually expected that knocking out Sun1 in these mouse models would worsen their conditions and cause them to die faster but surprisingly we observed the opposite," said IMB graduate student and co-author Rafidah Abdul Mutalif.

As mutations in Lamin A are frequently reported in hereditary cardiomyopathies, the scientists hope that their research findings may lead to a potential therapy for other forms of heart disease.

The article can be found at: Chen CY et al. (2012) Accumulation of the Inner Nuclear Envelope Protein Sun1 Is Pathogenic in Progeric and Dystrophic Laminopathies.

Source: A*STAR.
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