Mouse Model for Premature Aging: Zmpste24 Knockout Mice

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BACKGROUND

Progerias are rare genetic diseases characterized by premature aging including: retarded growth, osteoporosis, alopecia, and ultimately occlusive vascular disease. Many progeriod disorders are caused by mutations that lead to the accumulation of a lipid-modified form of prelamin A (farnesyl-prelamin A), resulting in a disruption of the cell nucleus. Zmpste24 is a mammalian integral membrane metalloproteinase that is critical for the processing of farnesylated proteins containing the carboxyl-terminal CAAX motif. Zmpste24, an ortholog of the yeast protein Ste24p, acts as an endoprotease by cleaving the 15 amino acids from the C terminus of prelamin A (including the farnesyl group), releasing mature lamin A.

INNOVATION

UCLA researchers have developed Zmpste24 knockout mice in order to determine the developmental and biochemical roll of Zmpste24. Zmpste24 -/- mice have retarded growth, hair loss, muscle weakness, spontaneous bone fracture, and shortened life spans. Zmpste24 deficient cells demonstrate the accumulation of wildtype farnesyl-prelamin A along the nuclear envelope, leading to misshapen nuclei. Researchers continue to utilize the Zmpste24 knockout mice to further understand the mechanism of progerias and ultimately provide treatment options for patients.

RELATED MATERIALS

- A protein farnesyltransferase inhibitor ameliorates disease in a mouse model of progeria. Science 2006.
- Zmpste24 deficiency in mice causes spontaneous bone fractures, muscle weakness, and a prelamin A processing defect. PNAS 2002.
- Biochemical studies of Zmpste24-deficient mice. J Biol Chem. (2001)

OTHER INFORMATION

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CONTACT

UCLA Technology Development Group
ncd@tdg.ucla.edu
tel: 310.794.0558.

INVENTORS

- Young, Stephen G.

OTHER INFORMATION

KEYWORDS

compliant electrode, dielectric elastomers, self-clearing, self-healing, dielectric breakdown, discharge, artificial muscles, mechanical actuators, solution processable metal.

CATEGORIZED AS

- **Medical**
  - Disease: Genetic Diseases and Dysmorphic Syndromes
  - Research Tools
- **Research Tools**
  - Animal Models

RELATED CASES

2007-222-0

ADDITIONAL TECHNOLOGIES BY THESE INVENTORS

- A New Mechanism For Hypertriglyceridemia In Humans
- Monoclonal Antibodies Against GPIHBP1
- Monoclonal Antibodies Against Prelamin A