




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Metabolic Brain Disease

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## FOXRED1 silencing in mice: a possible animal model for Leigh syndrome (Article)

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### Abstract

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Leigh syndrome (LS) is one of the most puzzling mitochondrial disorders, which is also known as subacute necrotizing encephalopathy. It has an incidence of 1 in 77,000 live births worldwide with poor prognosis. Currently, there is a poor understanding of the underlying pathophysiological mechanisms of the disease without any available effective treatment. Hence, the inevitability for developing suitable animal and cellular models needed for the development of successful new therapeutic modalities. In this short report, we blocked FOXRED1 gene with small interfering RNA (siRNA) using C57bl/6 mice. Results showed neurobehavioral changes in the injected mice along with parallel degeneration in corpus striatum and sparing of the substantia nigra similar to what happen in Leigh syndrome cases. FOXRED1 blockage could serve as a new animal model for Leigh syndrome due to defective CI, which echoes damage to corpus striatum and affection of the central dopaminergic system in this disease. Further preclinical studies are required to validate this model. © 2018, Springer Science+Business Media, LLC, part of Springer Nature.

### SciVal Topic Prominence

Topic: Mitochondrial Diseases | Electron Transport Complex I | assembly factor

Prominence percentile: 95.162 

### Author keywords

[FOXRED1](#) [Gene silencing](#) [Leigh syndrome](#) [Neurodegenerative diseases](#)

### Indexed keywords

EMTREE drug terms: [foxred1 protein](#) [oxidoreductase](#) [small interfering RNA](#) [unclassified drug](#)

EMTREE medical terms:

[animal experiment](#) [animal model](#) [animal tissue](#) [Article](#) [behavior change](#) [controlled study](#) [corpus striatum](#) [disorders of mitochondrial functions](#) [dopaminergic system](#) [gene silencing](#) [Leigh disease](#) [mouse](#) [mRNA expression level](#) [nerve degeneration](#) [nonhuman](#) [open field test](#) [pathogenesis](#) [substantia nigra](#) [tremor](#)**Metrics**  [View all metrics](#) 

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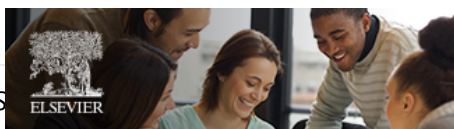
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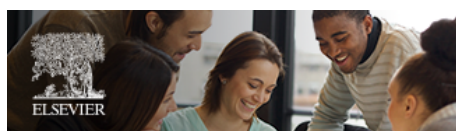
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