FOXRED1 silencing in mice: a possible animal model for Leigh syndrome

By: Salama, M (Salama, Mohamed)[1,2]; El-Desouky, S (El-Desouky, Sara)[1]; Alsayed, A (Alsayed, Aziza)[1]; El-Hussiny, M (El-Hussiny, Mahmoud)[1]; Moustafa, A (Moustafa, Abdelrahman)[1]; Taalab, Y (Taalab, Yasmeen)[2]; Mohamed, W (Mohamed, Wael)[3,4]

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Abstract

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.

Keywords

Author Keywords: FOXRED1; Neurodegenerative diseases; Leigh syndrome; Gene silencing

KeyWords Plus: MITOCHONDRIAL COMPLEX-I; FAD-DEPENDENT OXIDOREDUCTASE; ASSEMBLY FACTORS; MOUSE MODEL; DEFICIENCY; DISEASES; PATTERN; NDUFS4

Author Information

Reprint Address: Salama, M (reprint author)

+ Mansoura Univ, Fac Med, MERC, Mansoura, Egypt.

Reprint Address: Salama, M (reprint author)

+ Mansoura Univ, Dept Toxicol, Fac Med, Mansoura, Egypt.

Addresses:

+ [1] Mansoura Univ, Fac Med, MERC, Mansoura, Egypt

+ [2] Mansoura Univ, Dept Toxicol, Fac Med, Mansoura, Egypt

+ [3] Menoufia Univ, Fac Med, Dept Clin Pharmacol, Shibin Al Kawm, Egypt

+ [4] Int Islamic Univ, Kulliyyah Med, Dept Basic Med Sci, Kuantan, Pahang, Malaysia

E-mail Addresses: toxicsalama@hotmail.com

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