Clin Neurophysiol. 2019 Aug 16;130(10):1988-1994. doi: 10.1016/j.clinph.2019.07.029. [Epub ahead of print]

Improvement of motor conduction velocity in hereditary neuropathy of LAMA2-CMD dy^{2J}/dy^{2J} mouse model by glatiramer acetate.

Rabie M[1], Yanay N[2], Fellig Y[3], Konikov-Rozenman J[2], Nevo Y[4].

Author information

1 Institute of Neurology, Schneider Children's Medical Center of Israel, Tel-Aviv University, 14 Kaplan Street, Petach Tikva 49202, Israel; Pediatric Neuromuscular Laboratory, Felsenstein Medical Research Center, Tel-Aviv University, 14 Kaplan Street, Petach Tikva 49202, Israel.

2 Pediatric Neuromuscular Laboratory, Felsenstein Medical Research Center, Tel-Aviv University, 14 Kaplan Street, Petach Tikva 49202, Israel.

3 Department of Pathology, Hadassah-Hebrew-University-Medical-Center, Kiryat Hadassah P.O.B. 12000, Jerusalem 91120, Israel.

4 Institute of Neurology, Schneider Children's Medical Center of Israel, Tel-Aviv University, 14 Kaplan Street, Petach Tikva 49202, Israel; Pediatric Neuromuscular Laboratory, Felsenstein Medical Research Center, Tel-Aviv University, 14 Kaplan Street, Petach Tikva 49202, Israel. Electronic address: yoramne@clalit.org.il.

Abstract

OBJECTIVE: Glatiramer acetate (GA), an agent modulating the immune system, has been shown to cause significantly improved mobility and hind limb muscle strength in the dy^{2J}/dy^{2J} mouse model for LAMA2-congenital muscular dystrophy (LAMA2-CMD). In view of these findings and the prominent peripheral nervous system involvement in this laminin-α2 disorder we evaluated GA's effect on dy^{2J}/dy^{2J} motor nerve conduction electrophysiologically.

METHODS: Left sciatic-tibial motor nerve conduction studies were performed on wild type (WT) mice (n=10), control dy^{2J}/dy^{2J} mice (n=11), and GA treated dy^{2J}/dy^{2J} mice (n=10) at 18weeks of age.

RESULTS: Control dy^{2J}/dy^{2J} mice average velocities (34.49±2.15m/s) were significantly slower than WT (62.57±2.23m/s; p<0.0005), confirming the clinical observation of hindlimb paresis in dy^{2J}/dy^{2J} mice attributed to peripheral neuropathy. GA treated dy^{2J}/dy^{2J} mice showed significantly

improved average sciatic-tibial motor nerve conduction velocity versus control dy^{2J}/dy^{2J} (50.35±2.9m/s; $p<0.0005$).

CONCLUSION: In this study we show for the first time improvement in motor nerve conduction velocity of LAMA2-CMD dy^{2J}/dy^{2J} mouse model's hereditary peripheral neuropathy following GA treatment.

SIGNIFICANCE: This study suggests a possible therapeutic effect of glatiramer acetate on hereditary peripheral neuropathy in this laminin-α2 disorder.

KEYWORDS: Glatiramer acetate; LAMA2-congenital muscular dystrophy; Nerve conduction; Peripheral neuropathy; dy(2J)/dy(2J) mice

PMID: 31476705 DOI: 10.1016/j.clinph.2019.07.029

LinkOut - more resources