Improvement of motor conduction velocity in hereditary neuropathy of LAMA2-CMD dy²J/dy²J mouse model by glatiramer acetate.

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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 dy²J/dy²J 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 dy²J/dy²J motor nerve conduction electrophysiologically.

METHODS: Left sciatic-tibial motor nerve conduction studies were performed on wild type (WT) mice (n=10), control dy²J/dy²J mice (n=11), and GA treated dy²J/dy²J mice (n=10) at 18weeks of age.

RESULTS: Control dy²J/dy²J 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 dy²J/dy²J mice attributed to peripheral neuropathy. GA treated dy²J/dy²J 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.

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


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