Myostatin inhibition using mRK35 produces skeletal muscle growth and tubular aggregate formation in wild type and TgACTA1D286G nemaline myopathy mice.


Abstract
Nemaline myopathy (NM) is a heterogeneous congenital skeletal muscle disease with cytoplasmic rod-like structures (nemaline bodies) in muscle tissue. While weakness in NM is related to contractile abnormalities, myofiber smallness is an additional abnormality in NM that may be treatable. We evaluated the effects of mRK35 (a myostatin inhibitor developed by Pfizer) treatment in the TgACTA1D286G mouse model of NM. mRK35 induced skeletal muscle growth that led to significant increases in animal bodyweight, forelimb grip strength and muscle fiber force, although it should be noted that animal weight and forelimb grip strength in untreated TgACTA1D286G mice was not different from controls. Treatment was also associated with an increase in the number of tubular aggregates found in skeletal muscle. These findings suggest that myostatin inhibition may be useful in promoting muscle growth and strength in Acta1-mutant muscle, while also further establishing the relationship between low levels of myostatin and tubular aggregate formation.

PMID: 29293963 DOI: 10.1093/hmg/ddx431
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