The Role of a Novel Integrin Enhancing Protein Therapy to Protect Skeletal Muscle from Damage and Its Relationship to Fiber Type in a Mouse Model of Duchenne Muscular Dystrophy

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There are more than 30 forms of Muscular Dystrophy which combined affects nearly 20,000 children born annually worldwide. Duchenne Muscular Dystrophy (DMD) is the most common form of muscular dystrophy. An inherited X-linked recessive disorder in which the protein dystrophin is no longer produced resulting in a lost connection from the cytoskeleton to the extracellular matrix causing structural weakness. Over time without dystrophin damage occurs in myofibers impairing normal muscle function. At this time there are no current cures for this disease. Skeletal muscle is made of two main fiber types, slow twitch (type 1a) and fast twitch (type 2a, 2b, and 2x) allowing for different actions and capabilities. Muscle can alternate between fiber types for a steadier supply of energy and may change in size or fiber type depending on the demands. This study hypothesized that a novel Integrin Enhancing Protein therapy (IEP) on skeletal muscle would decrease the amount of muscle damage. The findings suggest that the mdx mice treated with a novel Integrin Enhancing Protein had a decrease in centrally located nuclei compared to control treated mice which indicates less damage and need for repair in treated mice. It was also determine that there was a difference in specific fiber type distribution in type 2a fibers. Therefore data showed a relationship between fiber type distribution and disease progression in the mdx mouse model of DMD.