Fiber-type Switching in Osteogenesis Imperfecta Mouse Models

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Osteogenesis imperfecta (OI) is an autosomal dominant disease that causes short stature, bone fragility, frequent bone fractures, and muscle weakness. Approximately 85% of OI cases are caused by mutations in type 1 collagen chains (COL1A1 and COL1A2). Our lab uses the osteogenesis imperfecta murine (*oim*) model of OI in mice in which the homozygous *oim/oim* mice display type III severe OI. Muscle force deficits independent of muscle size have been found in patients and mouse models of OI, showing the inherent nature of muscle weakness in OI. Mitochondrial dysfunction has also been observed in the skeletal muscle of OI mouse models. Because mitochondria supply energy to muscles, we analyzed muscle fiber-types in OI and wild type mice to determine if OI can cause muscle fiber-type switching. Muscle fiber-types include slow-twitch, oxidative, type I fibers; type IIa fibers that use a mix of glycolytic and oxidative phosphorylation; and fast-twitch type IIb fibers that are the most glycolytic. Using immunohistochemical staining of myosin heavy chains, we stained type I, type IIa, and type IIb muscle fiber types. In this experiment, it was determined that homozygous *oim/oim* mice had significantly less type I fibers compared to wild type mice and that *oim/oim* mice had significantly more type IIa fibers than wild type mice.