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Muscle Fiber-typing in the G610C Mouse Model of Osteogenesis Imperfecta

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Osteogenesis Imperfecta (OI), also known as "brittle bone disease" is an autosomal dominant disease that causes short stature, bone fragility, frequent bone fractures, reduced bone mineral density, and inherent muscle weakness. Currently, the most common treatments for OI are surgical rodding and bisphosphonates. Because bisphosphonates inhibit bone remodeling, there is more bone, but it is poorer quality, therefore it is not a good treatment for children. An alternative way to increase bone strength is to increase muscle mass. When muscles pull on bone with more force, the bone becomes stronger in response. Muscles in patients and mouse models of OI have shown to have force deficits compared to wild type muscles. In a previous experiment, mice modeling severe OI were shown to have significantly less type I oxidative fibers compared to wild type. Mice with the same severe form of OI also display mitochondrial dysfunction in muscle tissues, which together may cause the muscle weakness. One target to make larger muscles is myostatin, a negative regulator of muscle mass. If there is less myostatin, muscles become larger. Using anti-myostatin antibodies, the circulating myostatin levels can be decreased, and muscles can grow larger. Mice with a mild to moderate form of OI, *G610C*, were injected with the anti-myostatin antibodies to determine if bone and muscle quality or size was improved. This project focuses muscle fiber-type composition changes between wild type and *G610C* mice and between mice of both genetic backgrounds given a control treatment versus the anti-myostatin antibodies. The results of this study will be shown at the Summer Undergraduate Research Forum. This project was funded by the generosity of Peggy and Andrew Cherng through the Cherng Summer Scholars Program.