Myotonia Congenita and Thyroid Function

Myotonia congenita (MC) is an ion channel disorder affecting the balance of electrolytes in the skeletal muscles caused by reduced function of the chloride channel, CLCN1. The condition is usually diagnosed by neurologists through a combination of genetic and clinical testing, and it's treated with various medications that can help balance the influx of positive and negative ions, primarily by blocking sodium channels and reducing the action potential of the muscle so that it can relax normally.

There have been numerous reports in scientific literature regarding increased myotonia in patients with hypothyroidism. This can occur even in patients without the inherited gene mutations that cause Thomsen's and Becker's myotonia congenita. Unlike myotonic muscular dystrophy where there are actual endocrine abnormalities, the increased stiffness caused by even subclinical hypothyroidism in MC seems to be more related to slight changes in the activity of the protein that controls the gating of the chloride channel.

There is a well-documented symptom called the warm-up phenomenon with myotonia congenita. When a person begins to move after a period of inactivity the muscles are extremely stiff until they are flexed several times. There is some speculation that the sodium-potassium pump is involved in this as well which is also temperature sensitive. Cold exposure exacerbates myotonia and warmth helps reduce the symptoms.

When I was diagnosed with myotonia congenita about forty years ago, my neurologist was an older gentleman who told me he always put his patients on low dose thyroid medication even if their labs were in normal range because it improved myotonia so much. I began with 1 grain of natural dessicated thyroid back then and saw quite a dramatic improvement.

Years later after experiencing significant muscle injuries when a doctor refused to believe there was a correlation and refused to fill a prescription, I sought an endocrinologist to help me sort it out. My TSH was within normal range, but he had worked with similar patients and said I needed to keep my TSH at .5 to 1 in order to get the best results. After monitoring free T4 and T3 as well as TSH we settled on 90 mg of Armour thyroid. I was fine with Thyrolar years ago which was synthetic T4 and T3, but whenever I tried to go back to T4 alone, the myotonia became much worse again. We have since learned that T3 is the important component for improving muscle function. It is often sufficient to use a timed release T3 but it has to be compounded which is a cost consideration since insurance will not cover that.

Many patients have confirmed improvement in their symptoms by using enough replacement to maintain TSH in the .5 to 1 range. I find that the stiffness increases noticeably when it gets to 2-3 which increases falls and muscle injuries. I know there is some concern about osteoporosis risk with long-term dosing that maintains a low normal level. However I am sixty-five and have used thyroid replacement for forty years. My recent DEXA scan shows normal bone density...not even osteopenia.

Obviously there are other medical conditions that can affect thyroid function, and hyperthyroidism can also negatively impact muscle function. But in general, we have found that maintaining a higher metabolic rate is beneficial for our disease and may be worth a trial to see if there is improvement.

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The Myotonia Project