CARPAL TUNNEL SYNDROME AND TRIGGER DIGITS

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Hand function can be severely impaired in children with MPS and ML, even after transplant or with Enzyme Replacement Therapy (ERT). Carpal tunnel syndrome, contractures of the fingers, and trigger digits are all common in children with MPS. *Flexion contractures* refer to the inability of joints to straighten out all the way, similar to the knees. This problem results from continued deposits of storage material in the soft tissues around the joints.

Trigger digits are fingers that sometimes stick in a bent position, but with continued prompting, straighten. Trigger digits are caused by deposits of storage material around the tendons. Tendons are the connections between muscles and bones responsible for actually moving the fingers. With continued deposition of mucopolysaccharides (LINK TO GLOSSARY), the tendon can get stuck in the pulley system, preventing full motion of the fingers. If the tendon pops through the pulley, it "triggers", thus the name "trigger digits".

Carpal tunnel syndrome results from continued deposits that compress the *median nerve* in the wrist. This nerve is responsible for some parts of hand sensation, and hand strength. Early symptoms for carpal tunnel syndrome may include pain, tingling, and numbness of the hand. These complaints are actually uncommon, however, in children with MPS and ML, because they are young and have difficulty communicating their symptoms, or they just become accustomed to the symptoms, not realizing they are abnormal. More commonly, parents notice that their children have increasing difficulty with fine motor tasks (such as handling a pen or pencil, using eating utensils, cutting with scissors, etc.) and decreased grip strength. The best way to diagnose carpal tunnel syndrome is by nerve studies, an electromyogram and nerve conduction velocities (EMG/NCV). It is now strongly recommended that children with MPS be tested yearly for carpal tunnel syndrome for early detection. Again, children won't typically complain about the symptoms, and the muscle weakness eventually noticed by parents is a late sign of nerve compression and consequently results in a poorer outcome with treatment.

Non-surgical treatment of hand problems includes regular visits with an occupational or physical therapist for evaluation and treatment. This should be started early, with the intensity of follow-up being individualized to each child's needs. As a general rule, occupational therapists are more specifically trained in the treatment of hand disorders, with some actually being "certified hand therapists". Among other things, they evaluate motor and sensory function, do range of motion exercises, practice fine motor tasks, and if necessary, make splints to help the symptoms of carpal tunnel syndrome.

Unfortunately, surgery is commonly required to treat these problems in children with MPS. Frequently, both hands are involved and can be treated at the same time. Trigger digits are treated by opening one or two of the many pulleys in each finger, to make more room for the tendon, and the tendon is itself cleaned of MPS deposits. Carpal tunnel syndrome is treated by releasing the constricting tissue over the nerve and removing the deposits on the surrounding tendons. These surgeries are relatively minor, but their benefit can be tremendous.