National MPS Society
Support for Families. Research for a Cure.

MPS: PAIN AND TREATMENT OPTIONS

Mucopolysaccharidoses are genetic diseases affecting 1 in 25,000 children. MPS and related diseases affect virtually every organ and body system including joints, nerves and muscles. This is due to incompletely broken down glycosaminoglycan (GAG) that remains stored inside the cells in the body and begins to build up, causing progressive damage and, for many, significant pain.

Individuals with MPS may present with symptoms of pain not unlike those with other degenerative diseases, including arthritis. This pain may occur from nerve pain stemming from neurologic involvement and neuropathic signals arising in the brain. There may also be joint pain that causes inflammation, stiffness and swelling in the knees, hips, hands and spine. Certain challenges in identifying the source or location of pain occur when working with non-verbal individuals. Joint pain may be noticed in non-verbal children/adults as weakness when holding something or falling down more often when standing. Another source of pain may be headaches such as migraines, increased intracranial pressure and/or tension. Non-verbal children and adults may manifest headaches by holding their head in their hands or not wanting to open their eyes, as well as crying when they are lying flat.

Related to the joint pain and neuropathic involvement, individuals and parents need to be aware of the possibility of spinal cord entrapment, compression and degeneration which can occur in MPS I, II, IV and VI. This can result in burning, numbness, tingling, and weakness. These symptoms in a non-verbal child may present as chewing on the hands or leg shaking.

There are a variety of pain management interventions that have been used with varying degrees of success including medications, supplements, massage, acupuncture and chiropractic care.

Pain control medications are broken down by class and selected based on the specific pain symptom being treated and the ability of the individual to tolerate medication side effects.

Over the counter oral medications such as aspirin and ibuprofen are usually the first lines of defense in treating pain. They work by reducing inflammation and reducing the pain associated with headaches. Prescription medications such as Darvocet, Percocet (Tylenol combination), and Celebrex are also commonly used. Anti-inflammatory medications can also be administered through a patch (Diclofenac) or gel (Voltaren).

Corticosteroids are a group of powerful anti-inflammatory medications which should be used with care due to the potential risk for gastro-intestinal and bone complications.

These are taken either orally (Prednisone) or injected directly into the joint or soft tissue (such as the spine) to reduce inflammation and swelling. Anti-depressants, anxiolytics and anticonvulsant medications have been used in the management of pain and adjunctively to address mood symptoms associated with chronic pain. Commonly prescribed medications include Cymbalta (*antidepressant*), Elavil (antidepressant) and Ativan (benzodiazepine). These medications may work by increasing the neurotransmitters in the spinal cord that reduce pain signals to the brain.

While widely used in pain management, it continues to be unclear exactly why the antidepressants reduce pain symptoms. Additionally, antidepressant medications have provided relief indirectly due to the sedating properties that assist individuals with sleep and mood as well as pain relief.

Medications such as Neurontin and Lyrica have proven to be successful in pain reduction for some individuals. They work by calming damaged or overactive nerves that cause pain. A Lidoderm patch, which is a topical anesthetic, is another treatment to help nerve pain.

Less often used but also beneficial, are a class of medications called opioids such as Fentanyl, Morphine and Dilaudid. These medications need to be used with caution and under the close supervision of the prescribing doctor. They are typically used in individuals who experience moderate to severe pain and have not experienced pain relief using other methods. It is important with these medications (oral, patch or gel) to work closely with your doctor and pharmacist when deciding whether to use a brand name or generic alternative. Some individuals have reported an increase or decrease in effectiveness of the medication when comparing the pain relief experienced using brand name or generic. Side effects to be aware of include constipation, dizziness, headaches, vision problems and cognitive changes.

Other pain control options include local anesthetics such as the Lidoderm patch. This has been found to work most often with neuropathic (nerve) pain. The patch is generally worn for 12 hours, followed by 12 hours off.

Muscle relaxants, such as Flexeril, Soma, Baclofen, and Skelaxin, may be used for spinal cord related muscle spasms. The intent of muscle relaxants is to help 'relax' the muscles, and to improve and relieve spasticity. Flexeril is used commonly in individuals with pain but may cause drowsiness, dizziness and lethargy.

Less commonly used but potentially effective are over-the-counter supplements such as vitamin B12 (methylcobalamin) for nerve pain, calcium and magnesium for joints, hyaluronic acid for joint problems, Vitamin D for bone related health and even Boswelia and Curamin which also can be used for bone health and pain. Physician consultation is necessary before beginning a supplement, to avoid potential serious interactions with existing medications.

Some of the medications used to treat and manage pain are regulated by the government (controlled). This is due to the risk associated with long term use and the high potential for abuse of these medications. In addition, with long term use there is the possibility of developing tolerance to the medication which would require increasing the dose to obtain the same pain relief previously experienced and/or physiologic dependence. Careful observation and close collaboration with a pain specialist or other physician is required.

Acupuncture and Chiropractic care are becoming more widely used by the general public for pain control. Acupuncture stimulates the release of pain-relieving endorphins in the body; influences the release of neurotransmitters, substances that transmit nerve impulses to the brain; influences the autonomic nervous system; stimulates circulation; and influences the electrical currents of the body.

The chiropractic profession treats muscle and joint pain through manual therapy that can

include manipulation of the spine, joints and soft tissue. Chiropractic can offer therapies and exercises beyond manipulation which may reduce tension, muscle and joint pain as well as neuropathic pain. Chiropractic and traditional medicine are often at odds about treatment of pain, but medical doctors and chiropractors are beginning to work hand in hand as a team approach. Physician consultation is strongly recommended before beginning chiropractic care, which may be contraindicated for some individuals with an MPS disease.

Physical and Occupational therapies may help reduce pain and improve overall functioning within the individual's environment, such as home and school. A TENS (Transcutaneous Electrical Nerve Stimulation) Unit is a small, battery-operated devise that sends electrical impulses to block pain signals from an area of the body. A physical or occupational therapist can prescribe TENS Unit.

Summary

It is important to talk with your physician about treatment options and what is recommended for you or your child. Not all individuals with an MPS disease have the same benefit or reaction from a specific treatment and some treatments may be contraindicated for specific MPS diseases. If a treatment is not beneficial, it should be stopped and another considered. The National MPS Society does not endorse specific treatments.