

TREATMENT SECTION

Word Count: 2,538

Date: 1/15/03

KEY TO HEADINGS

A-level: **PHYSICAL THERAPY**

B-level: **Physical therapy**

C-level: **Physical therapy**

Text:

Treatment is available for some of the symptoms experienced by those with HSP or PLS. While there is currently no cure and no treatment that can prevent, slow or reverse the progression of these similar neurological disorders, symptomatic treatment can help minimize the physical effects of HSP and PLS caused by degeneration of the primary upper motor neurons. Both conditions are treated alike, except in the case of voice/swallowing problems, which are associated with PLS and not HSP.

Spasticity (increased muscle tone, stiffness), muscle weakness, abnormal gait, decreased sense of balance, urinary and bowel problems, shortened Achilles tendons and nighttime leg cramps can all be reduced or dealt with through physical therapy, exercise, drug therapy, surgery, orthotics and assistive devices. Generally, a combination of these methods can be expected to provide the best problem management. Certain food supplements and alternative treatments may also be worth your interest. As with any medical condition, it is important to focus on what you can do to help yourself and join the patient community to discover the support and advice of others.

PHYSICAL THERAPY AND EXERCISE

Physical therapy, or a regular stretching and exercise program, can be effective in reducing a range of symptoms, both direct and indirect. It will help maintain mobility and range of motion and can help retain or improve muscle strength. Also, it can minimize muscle atrophy, increase endurance, reduce fatigue, reduce the occurrence of spasms and cramps, and provide cardiovascular conditioning. As a bonus, the natural positive psychological effects of exercise help to lessen stress and produce feelings of well-being.

A doctor or physical therapist should tailor your exercise program, as patients' needs vary. Some patients have severe spasticity but only mild weakness, some have more weakness than spasticity, and others experience the two more or less equally. Healthcare professionals will use various tests to determine the degree of spasticity and recommend specific exercises to accommodate your needs and capabilities. A prescribed program may include stretching, aerobic and strengthening activities.

Stretching Exercises

Stretching will help maintain or increase range of motion (flexibility) and reduce muscle spasticity and cramps. The rate of problems such as tendonitis and bursitis is also reduced through stretching.

Aerobic Exercises

Aerobic exercises are important even for people without HSP or PLS. Walking, bicycling, swimming and water aerobics are excellent activities in this category. They improve cardiovascular and general fitness, reduce fatigue and increase endurance. For those with limited use of their legs, upper-body exercises can provide these benefits.

Strengthening Exercises

Strengthening exercises help build up the muscles that have not yet weakened and slow the rate of impairment. Exercise to strengthen often-unused muscles will minimize atrophy, such as may occur in the calves of patients who use wheelchairs. To achieve strengthening, weight training or aquatic exercises may be recommended. And exercises for back muscles may eliminate much of the pain resulting from poor gait, poor posture or the use of mobility aids.

While stretching and aerobics are consistently recommended, not all doctors will prescribe, and not all patients will embrace, a strengthening routine. Some patients are not affected with muscle weakness, and others may not judge as worthwhile the time and effort necessary to see small improvements. All strength training requires patience and significant amounts of time! And while weakness may be slowed by maintaining a strengthening program, rejecting mobility aids, or both, you may find an easier or fuller life possible if you do not worry about keeping up muscle strength. It depends on what is important to you.

Spasticity can limit the ability to exercise muscles. Antispasmodic drugs may help improve the results of physical therapy or exercise.

DRUG THERAPY

Medication is prescribed to people with HSP or PLS mainly to treat symptoms of spasticity (including cramps) and urinary/bowel problems. Some drugs (such as Tizanidine) can occasionally cause liver damage, so it may be important to have your liver functions checked periodically.

For spasticity

Various drugs (antispasmodics) are used to combat the spasticity associated with HSP and PLS (and other disorders, such as multiple sclerosis). Spasticity may range, in different patients, from mild to severe. Antispasmodics are often used in conjunction with physical therapy and can reduce spasms, stiffness and cramps.

With any of the following antispasmodics, it is important to not reduce your dosage or suddenly stop without consulting your doctor. Some (especially baclofen) can cause serious problems such as seizures or hallucinations if your dosage is not correctly tapered. If you feel you need to reduce or stop your medication, talk to your doctor!

Baclofen

Brand name: Lioresal. Outside the United States: Alpha-Baclofen, Baclon, Baclosal, Baclospas, Baklofen, Clofen, Lebic, Mulax, Pacifen, Spinax

Baclofen is a muscle relaxant and antispasmodic agent. Another benefit of baclofen is that it may relieve urinary problems, though other drugs are better suited to this purpose (see For urinary/bowel problems below).

Baclofen may be taken orally. It also may be injected via an intrathecal baclofen pump, surgically implanted in the abdomen to deliver small, continuous doses of medication directly to the spinal canal for better performance and fewer side effects. Intrathecal baclofen has traditionally been used for cerebral palsy and people with brain or spinal-cord injury who have severe spasticity. However, it can also be helpful to those with less severe spasticity.

If you are taking oral baclofen and considering the advantages of an intrathecal pump, prior to implantation you will be given one or more test doses of baclofen by spinal injection. This allows the physician to find whether this method of delivery will work well for you and determine the appropriate dosage. Once the pump is implanted, there is no further need to puncture the skin until the pump is refilled, about every four months.

Some patients find that baclofen helps them walk better; others find no effect, or that the reduced spasticity exposes the weakness of the legs too much and actually increases walking difficulty.

Side effects of baclofen may include fatigue, memory disturbance, slurred speech and impaired coordination. If you take baclofen mainly to reduce nighttime spasms, talk to your doctor about adjusting your dosage to take it just at night, or a heavier dose at night, to decrease problems with drowsiness during the day.

Information from the manufacturer regarding Medtronic ITB (Intrathecal baclofen) Therapy can be found at www.medtronic.com.

Diazepam

Brand names: Valium, Diastat

Clonazepam

Brand name: Klonopin

Diazepam and Clonazepam are benzodiazepine agents. This family of drugs consists of sedatives that slow the central nervous system and are therefore useful in treating spasticity. There are many brand names for diazepam outside of the United States.

Side effects may include drowsiness and weakness.

Tizanidine

Brand name: Zanaflex. Outside the United States: Sirdalud, Sirdalud MR, Sirdalud Retard, Ternelax, Ternelin

Tizanidine is a short-acting drug, useful for treating nocturnal spasms. Its use should be individualized and directed at those times when relief of spasticity is most important.

There have only been limited studies of patients exposed to long-term use, so caution is advised. *If you take Tizanidine, it is important to have liver functions tested regularly.*

Dantrolene sodium

Brand names: Dantrium, Dantrium IV

Dantrolene sodium is less likely than the benzodiazepams to cause drowsiness or confusion, but it may cause general weakness.

It can cause liver damage, and so is not usually prescribed unless other antispasmodics have not helped. *It is very important to have liver enzymes and functions monitored while on Dantrolene sodium.*

Botulinum toxin

Brand name: Botox

or phenol treatment (chemodenervation)

Botox injections are generally considered only for rare cases with severe spasticity. Botulinum toxin is injected directly into the muscle to be treated, relaxing it. This then allows for range-of-motion exercises to help lengthen the muscles. The treatment must be repeated periodically.

Gabapentin

Brand name: Neurontin

Gabapentin, widely used to treat seizures and neuropathic pain (such as in people with MS or diabetes), is also useful in reducing spasticity, according to recent reports. It is usually well tolerated and may be considered as an option for those who experience too many side effects with baclofen.

For weakness

4-aminopyridine

4-aminopyridine (4-AP) is a blocker of potassium channels, used to help nerve conduction in demyelinated axons (branches of nerves stripped of their protective sheaths). It is an experimental drug, not yet approved by the FDA and thus not likely to be covered by medical insurance. 4-AP has been used with some success in treatment of MS-related fatigue, muscle weakness and heat sensitivity. At least one person with HSP has been taking it and reports improved strength.

For urinary/bowel problems

If you have HSP or PLS, you may experience increased urgency or frequency in urinary or bowel functions. It is worth noting, however, that these problems can also have causes other than HSP or PLS, so it is important to discuss such symptoms with your doctor. As well as the prescription medications listed below, there are some over-the-counter bulking agents that can help, such as psyllium (brand names: Metmucil, Konsyl)—ask your doctor about them.

Oxybutynin chloride

Brand name: Ditropan XL

Oxybutynin chloride is antispasmodic and anticholinergic (works against acetylcholine, a neurotransmitter chemical in muscles). It reduces bladder contraction and the urge to void. Oxybutynin chloride should be used sparingly, as it creates urinary retention. Its use is best reserved for periods when bathrooms will not be readily available, such as on long trips.

The most common side effect is dry mouth. Other side effects may include constipation, drowsiness, diarrhea, blurred vision, dry eyes, dizziness and runny nose.

Tolterodine tartrate

Brand name: Detrol

Detrol is similar in action to oxybutynin chloride.

Side effects may include dry mouth, headache, dry eyes, constipation and indigestion.

Dicyclomine

Brand name: Bentyl

Dicyclomine relieves spasms of the gastrointestinal tract (stomach and intestines) by blocking the actions of spasm-inducing chemicals in the body. It is used to treat functional bowel or irritable bowel syndrome (IBS) and bladder spasm.

Hyoscyamine

Brand names: Levsin, Anaspaz, Levsin

Levsin is an extended-release hyoscyamine sulfate and another anticholinergic. It often causes dry mouth and constipation. Blurred vision may be another side effect.

Since certain foods and drinks can bring on or aggravate urgency, it may be helpful to avoid them as possible. Some common substances are caffeine, alcohol, acidic fruits and juices, spicy foods and artificial sweeteners.

It may also help reduce urinary frequency to try “double voiding:” Urinate as much as possible, wait briefly, then attempt to urinate again.

SURGERY

Shortened Achilles tendons (heel cords) may occur in people with HSP or PLS. This is more common when symptoms begin in childhood and there has not been sufficient physical therapy.

Surgery to lengthen the tendons may be recommended in some cases, although results may not be permanent.

ORTHOTICS

The effects of HSP and PLS can sometimes make walking difficult—orthotics can provide some physical correction. They are special shoe inserts, splints or braces used to relieve various gait problems, relieve foot problems, help increase balance or remove pressure from sore spots.

You may find preformed devices like simple arch supports available at drugstores. However, such over-the-counter orthotics may not be very effective—it is advised that you consult a physical therapist for specific recommendations and obtain custom-made devices fitted by professionals.

Besides orthotics, choice of shoes can also make a difference for your ease and comfort of walking. Styles with toes that curve up (running shoes) can help reduce tripping and toe wear. High-top shoes or boots can provide extra ankle support for a steadier gait. Sturdy soles, or repairing worn areas of soles, may also help.

ASSISTIVE DEVICES

A cane or other mobility aid can help compensate for the decreased sense of balance common in HSP or PLS. Not only will it provide you more stability, it will help avoid fatigue from overexertion.

Almost everyone resists the idea of needing an assistive device, for any of various reasons. Yet as a rule, those who choose to use one find it much easier and claim to notice *less* attention from others than they were subject to while they walked unsupported (for those self-conscious).

A little searching should yield an attractive cane or walking stick to suit your style. Or, spruce up an unremarkable one yourself with a little imagination.

Extra wary of winter? Freeze at the thought of walking across icy patches? For added traction, a cane tip with flip-up spikes or special grips can replace the ordinary tip.