Key Points

1. Neurological exam can identify whether a syrinx is causing problems with sensory nerves
2. Tests involve activating specific nerves electrically or with lasers and measuring the response
3. Study looked at 6 SM patients with MRI verified syrinxes and suspected of having areas of sensory deficit
4. Patients were given temperature, pain, and evoked response tests
5. Electrically evoked responses were normal, but laser evoked responses in the same area were clearly abnormal
6. Specific type of test, sympathetic sudomotor skin test were also abnormal
7. Type of test used in evaluating sensory deficits with SM may be important

Definitions

dermatome - an area of the skin associated with a specific nerve root

laser evoked potential (LEP) - test of the somatosensory nerves which uses a laser as stimulus and measures the time of response

somatosensory - sensory activity involving skin, muscles and organs

somatosensory evoked potential (SEP) - test which uses electrical signals to stimulate specific nerves and measures the time it take the nerve signals to travel

sympathetic nervous system - nerves, which are not consciously controlled, involved in preparing the body for physical activity

eSSR - electrical sympathetic

Syringomyelia Disrupts Pain & Temperature Sensing

May 31, 2007 -- Many Chiari and syringomyelia patients never undergo a thorough neurological exam, but for those who do, the multitude of probes and instruments used to prick, poke and prod can appear to be more at home in a low budget horror movie rather than a modern medical office. Despite their appearance, the tools used for a neurological exam can actually reveal quite a bit of information about neurological problems. For Chiari and syringomyelia patients, the neurological exam is an objective, critical complement to an MRI and can show whether the herniated tonsils and/or syrinx are causing specific problems with the nervous system and to what extent.

The neurological exam works because the human nervous system is highly organized. When you touch something, specialized receptor cells in your skin send electrical signals along nerve fibers to the brain. Similarly in order to move, the brain sends electrical signals to your muscles, which in turn send signals back to the brain so you have an internal sense of where your body parts are and how they are moving.

The nerve fibers that carry all this electrical messaging are laid out like a system of roads. Major bundles of nerve fibers, like highways, run down the spinal cord. At each spinal level, or segment, groups of nerve fibers branch out into the body like primary roads. As the nerves get closer to their final destination - like your shoulder or thumb - the nerve fibers branch out even more to serve specialized cells.

Like with any set of roads, a map can be used to help navigate the nervous system. Scientists have been investigating how people respond to stimuli for hundreds of years, but the neurological exam itself began to evolve in the late 1800's. The medical researchers of the time began to realize that different types of sensation - temperature, touch, pain - were affected differently by injury and disease, and traveled different pathways to the brain. Normal responses to stimulus were characterized, as were normal joint movements. As knowledge of the nervous system advanced, testing for sensation became more prevalent. By the 1950's, the neurological exam contained many of the features present in today's exam.

The guideposts along the human nervous system roadmap are the spinal segments from which nerve bundles branch out. The spine is composed of 7 cervical, 12 thoracic, 5 lumbar, and 5 sacral segments with the cervical segments at the top and the sacral segments at the bottom. Each spine segment is denoted by its region and number, C4 for example is the fourth segment down of the cervical region and L3 is the third segment down of the lumbar region. The nerves that branch out from the spine at each segment serve - or map to - a specific location in the body, called a dermatome. The cervical segments generally serve the neck and shoulders, the thoracic region maps to the chest, the lumbar region maps to the hips and the front of the legs, and the sacral segments map to the back of the legs and part of the feet.

What makes this mapping useful is the fact that damage to the nerve root will cause a loss of sensation in the area served by that nerve. So if doctors detect a loss of sensation, or muscle strength, in the shoulders, there is likely a problem at the C4 level.

In addition to mapping dermatomes, different types of stimulus can be used by doctors to aid diagnosis. Your skin and organs contain different types of receptors which specialize in responding to touch, pressure, pain, vibration, and temperature. The different types of receptor cells send their information along different sized nerve fibers - which mean the signals travel at different speeds - and along different routes.

Physicians can use this to isolate problems by activating different types of nerves and measuring the electrical response. One common test of this type is somatosensory evoked potentials (SEPs). The somatosensory nerves are nerves which send information from parts of the body, such as the skin, muscles and organs, other than the specialized sensory areas like the eyes and ears. SEPs work by exciting a specific nerve with an electrical signal and measuring how long the signal takes to travel a specific distance. If the signal is delayed, or does not get through at all, it is an indication of a spinal problem at a specific location. SEPs tend to be rather crude tests and a newer variation, LEPs, involve lasers to evoke the nerve response.

Similar to evoked potentials, in that either electricity or lasers can be used as a stimulus, SSRs (sympathetic sudomotor skin response) are a simpler test used to evaluate sympathetic nerves. Sympathetic nerves are a part of the nervous system which are not consciously controlled and prepare the body for physical action. Like SEPs, SSRs stimulate these nerves with electricity, laser or heat and measure their response time.

Even though syrinxes are well known to casues sensory problems, tests such as SEPs, LEPs and SSRs are not necessarily used to evaluate every syringomyelia patient. Many surgeons believe the fact that a syrinx exists is enough to justify surgery and may limit their neurological exams to simpler tests which do not involve lasers or electrical stimulation.

sudomotor skin response; test which uses electrical signals to stimulate sympathetic nerves and measures the response

ISSR - laser sympathetic sudomotor skin response; test which uses a laser to stimulate

cerebellar tonsils - portion of the cerebellum located at the bottom, so named because of their shape

cerebellum - part of the brain located at the bottom of the skull, near the opening to the spinal area; important for muscle control, movement, and balance

cerebrospinal fluid (CSF) - clear liquid in the brain and spinal cord, acts as a shock absorber

Chiari malformation I - condition where the cerebellar tonsils are displaced out of the skull area into the spinal area, causing compression of brain tissue and disruption of CSF flow

decompression surgery - general term used for any of several surgical techniques employed to create more space around a Chiari malformation and to relieve compression

Source

Veciana M, Valls-Sole J, Schestatsky P, Montero J, Casado V.
Abnormal sudomotor skin responses to temperature and pain stimuli in syringomyelia.
J Neurol. 2007 Apr 10; [Epub ahead of print]

However, a recent study from a group of neurologists in France (Veciana et al.) has shown that the type of neurological tests used to evaluate sensory deficits in syringomyelia may be important. Published on-line in the Journal of Neurology, the report detailed their experience in using a variety of tests to evaluate sensory problems in six syringomyelia patients.

Each patient had a syrinx that was clearly identifiable on MRI and was suspected of having at least one area of sensory loss (see Table 1). (Ed. Note: It is interesting that most of the patients had problems with their right side. I don't know if this has been studied rigorously, but I believe that for unknown reasons, problems on the right side are more common than the left in SM patients.)

Table 1:
Areas of Sensory Deficits In Six Syringomyelia Patients

Patient	Syrinx Location	Pain & Temp Sensory Loss
1	C1-conus	Right upper limb
2	C3-C4	Right upper and lower limbs
3	C2-T2	Right lateral neck
4	C3-T2	Right upper thoracic
5	T7-conus	Right lower limb
6	C2-T8	Left upper limb; abdomen; both lower limbs

As part of their exam, the area in question (for example right shoulder) for each patient was evaluated using a number of techniques: warmth threshold, cold threshold, heat pain, laser pinprick, SEPs, LEPs, eSSRs and lSSRs. The results were compared to the same section of skin on the other, and supposed normally functioning, side of the body.

Not surprisingly to anyone with a syrinx, the temperature and pinprick thresholds were higher in the affected areas for every patient than in the non-affected areas. In other words, the affected areas were not able to detect temperature and pricks as well as they should be able to. What was surprising was that the electrically stimulated tests, both SEPs and eSSRs did not produce abnormal results, whereas the laser stimulated ones (LEPs, lSSRs) produced clearly abnormal results indicating the syrinx was affecting those nerves (see Table 2).

Table 2
Sensory Results By Test Type

Test	Result
Warmth Threshold	Abnormal
Cold Threshold	Abnormal
Heat Pain	Abnormal
Pinprick	Abnormal
SEPs	Normal
LEPs	Abnormal
eSSR	Normal
lSSR	Abnormal
thermal SSR	Abnormal

SEPs are notoriously crude tests, but these findings bring into question their utility in evaluating sensory deficits associated with a syrinx when the areas being tested were clearly compromised yet the tests showed normal results.

The authors also point out that this is the first published report of the sympathetic sudomotor skin response tests being successfully used to identify and quantify syrinx damage to the sympathetic nervous system.

This is interesting because the sympathetic nervous system is involved in regulating many of the automatic functions of the body, such as sweating, pupil dilation, urine output, and hormone levels associated with the adrenal gland. Many syringomyelia patients have difficulty regulating their body temperature and experience abnormal sweating, which are signs of sympathetic nerve problems. Although the authors do not discuss this, it is also worth thinking about the fact that the sympathetic nervous system controls the fight or flight stress response and one has to wonder how this might manifest in what patients experience.

Given the relative simplicity of administering SSRs, it would seem to be worth considering as a standard test in evaluating syringomyelia patients to objectively determine both the effects of the syrinx and whether surgery can improve the sympathetic nerve problems.

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