Definitions

afferent - carrying a signal from the body to the brain

ataxia - inability to coordinate muscle movements, usually refers to walking in an abnormal way

biceps - large muscle at the front of the upper arm

clonus - rapid contraction and relaxation of a muscle, like a spasm

cranial - pertaining to the cranium, or skull

dermatome - an area of skin served by a spinal nerve

dysco-ordination - abnormal co-ordination

efferent - carrying a signal from the brain to the body

gait - act of walking

hamstrings - muscles in the back of the thighs

MRI - Magnetic Resonance Imaging, diagnostic device that uses a magnetic field to create an internal image of a person

proprioception - perception of movement and position from internal signals (not the eyes, etc.)

quadriceps - large muscles on the front of the thighs

reflex - involuntary response to a stimulus

spontaneous venous pulsations (SVP's) - periodic changes in size of the veins in the retina (in the back of the eye) observable in a large percentage of people

tendon - fibrous tissue that connects muscle with bone

trapezius - two large triangular muscles that run from the back of the skull to the middle of the back; used for raising the head and shoulders

triceps - muscles along the back of the upper arm

The Neurological Exam

"Hold your arms like this and resist."
"Do you feel this?"
"Walk on your heels across the room."
"Squeeze my fingers."

Anyone who has been diagnosed with CM/SM, or is even suspected of having a neurological problem, has gone through some variation of a neurological exam, probably more than once. The first time I was asked to perform some of the neuro tricks, I was surprised by what I couldn't do. Button a shirt with one hand - nope; walk on tip-toes - nope; stand still with my eyes closed - nope, I started to fall.

At the time, I didn't know what the tests meant, other than something was very wrong. As I learned later, the neurological exam is the most fundamental diagnostic tool that neurologists and neurosurgeons use to identify and isolate problems. By testing for strength, reflexes, reaction to touch and temperature, and a variety of other items, doctors are able to deduce where in the body the nervous system isn't working properly and how severe any damage to the nervous system is.

Anatomical Basis of the Neurological Exam

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 (more on spinal segments in a bit). 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 (right); C4 for example is the fourth segment down of the cervical region, 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. A different type of receptors, known as proprioceptive cells, provide information on movement and position. 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. So by combining responses to different stimulus, along with motor functions such as strength and gait, doctors can begin to get a picture of the location and extent of damage to the nervous system.

The Neurological Exam

A complete neurological exam is an extensive procedure which involves many tests. In addition to evaluating a patient's medical history and mental status, in general the tests can be grouped into the following categories:

Sources

- Freeman C, Okun MS. Origins of the Sensory Examination in Neurology. Seminars in Neurology 22(4); December, 2002.
- University of Florida web site
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- San Francisco University web site
- Spineuniverse.com

- Cranial Nerves
- Motor Function
- Coordination and Gait
- Reflexes
- Sensation

Because specific diseases present different neurological symptoms, a neurologist or neurosurgeon will usually tailor an exam to the specific patient and situation and focus on a few key areas. For example, according to Dr. Ghassan Bejjani, Clinical Assistant Professor of Neurological Surgery at the University of Pittsburgh, patients with a Chiari I malformation, but no syrinx, many times don't show any deficits with a neurological exam except for a finding involving the veins in their eyes. While the exam is still important for people with Chiari, for people with Chiari and a syrinx, "[The exam] is a way to assess whether the disease is causing any deficits warranting surgical intervention...[I look for] evidence of sensory deficits, weakness, ataxia, dysco-ordination, lower cranial dysfunction, and lack - or presence of - spontaneous venous pulsations." Dr. Bejjani goes on note that the neurological exam is a fairly objective test - the results won't vary much doctor to doctor - however which parts of the exam are performed depends on the situation, "The thoroughness [of the exam] depends on whether a syrinx is present, whether it is a follow-up or an initial visit, and how important the findings will be in the surgical decision making."

Cranial Nerves

These tests assess the nerve functions of the head, neck, and shoulders and might include tests involving the eyes, ears, nose, throat, jaw, neck muscles, and trapezius muscles. A doctor checking for CM/SM related deficits might pay particular attention to lack of neck movement, weakness in the trapezius muscles (the shoulder shrug muscles), hoarseness of voice, abnormal gag reflex, and as Dr. Bejjani mentions, spontaneous venous pulsations - SVP's.

In the vast majority of people, the veins in the retina in the back of the eye can be seen to pulsate when a doctor looks through the pupil. For people with elevated intracranial pressure - the pressure inside their head - these pulsations tend to not occur. Elevated intracranial pressure (ICP) can be associated with a Chiari malformation. So if a doctor observes the lack of SVP's, it is a good indication of high intracranial pressure and might indicate a Chiari malformation.

One of the classic symptoms of syringomyelia is pain and stiffness in the neck and shoulders, or what is known as the cape effect. If a doctor observes stiff neck movements and weak trapezius muscles, it might indicate a symptomatic syrinx.

Motor Function

When a doctor evaluates motor function, he is essentially checking whether the nerves that supply your muscles are working. A doctor will evaluate muscle size, tone (flaccid vs. rigid), and strength and look for weakness, imbalance between right and left sides, and muscles that are either too rigid or too soft.

Muscle strength is rated on a scale from 0-5 (below) and is measured by a patients ability to resist force. Basically, the patient tries to hold his arms (or legs) in a specific position while the doctor applies force. Evaluations might include tests for the biceps, triceps, wrist, hand, hip muscles, quadriceps, hamstrings, and ankles. People with syringomyelia tend to lose strength in at least one of their hands, and in many cases the hand will noticeably shrink in size over time.

Motor Strength
Assessment Scale (0-5)

Score	Description
0	No Muscle Movement
1	Some visible movement
2	Full range of motion, not against gravity
3	Movement against gravity, but not resistance
4	Movement against resistance, less than normal
5	Normal strength

Coordination and Gait

These are the tests that make you feel like you've been pulled over by the state police for DUI. The coordination and gait tests can reveal problems with the nerves that provide feedback on muscle movement and position, balance, and cerebellar function. Tests for coordination include:

- Rapid alternating movements such as touching your thumb to the tips of your fingers in succession.
- Point to point movements - the old touch something with your finger, then touch your nose (may be done with eyes closed)
- Romberg test - Named after the 19th century German neurologist who developed it, the Romberg test

involves standing with your feet together and eyes closed for about 10 seconds. If you lose your balance, it indicates a problem along one of the highways in the spinal cord.

Gait, or walking, requires many nerve functions to work together and is a good indication of problems in the nervous system. In addition to walking normally, a doctor may observe how a patient walks on their toes, walks on their heels, or walk heel-to-toe along a line. During my initial exam, while I was able to walk normally, I struggled to walk on my toes or heels and I failed the Romberg test by falling into the doctor.

Reflexes

The neurological exam tests what are called deep tendon reflexes. Basically, a doctor uses a small hammer to strike a tendon and watches the response. In many CM/SM cases, there will be an exaggerated response movement, indicating a problem - or lesion - of the muscle nerves in a specific location. As discussed earlier, the location of the problem can be deduced by identifying which reflexes are abnormal. The exam might include testing the following reflexes:

- Biceps - Correlates to C5, C6
- Triceps - Correlates to C6,C7
- Forearm - Correlates to C5,C6
- Abdomen - Correlates to T8-T12
- Knees - Correlates to L2-L4
- Ankles - Correlates to S1,S2

Reflexes are graded on scale from 0-4 (below), with 0 being no reflex, and 4 being an abnormally strong reflex with clonus - a series of muscle contractions.

Reflex Grading Scale (0-4+)

Score	Description
0	No Reflex
1+	Hypoactive (less than normal)
2+	Normal
3+	Hyperactive (more than normal)
4+	Hyperactive w/ Clonus (like a muscle spasm)

Sensation

As mentioned earlier, the sensations of touch, pain, temperature, vibration, pressure are carried along different nerves and pathways. By testing with different stimulus at different locations, a doctor can locate potential problems. A sensation exam might include using Q-tip to test for light touch, a tuning fork to test for vibration, and pin pricks. The exam will test sensation on the arms, legs, and other areas depending on the findings and patient history.

The Neurological Exam in the Age of MRI's

Before the development of advanced imaging technique like the MRI, which can clearly identify Chiari malformations and syrinxes, the neurological exam was the main diagnostic tool in the neurological arsenal. Despite rapid and impressive advances in imaging, the exam is still critical for CM/SM patients as it can identify the type and extent of any neurological deficits, important factors when evaluating treatment options.