



Syringomyelia

U.S. DEPARTMENT OF HEALTH
AND HUMAN SERVICES
National Institutes of Health

Syringomyelia

What is syringomyelia?

Syringomyelia is a disorder in which a fluid-filled cyst (called a syrinx) forms within the spinal cord. This syrinx can get bigger and elongate over time, damaging the spinal cord and compressing and injuring the nerve fibers that carry information to the brain and from the brain to the rest of the body.

A watery liquid known as cerebrospinal fluid (CSF) normally surrounds and protects the brain and spinal cord. CSF also fills connected cavities within the center of the brain called the ventricles which continue to a small central canal that runs the length of the spinal cord. When a person has syringomyelia this fluid collects within the tissue of the spinal cord, expands the central canal, and forms a syrinx. Generally, a syrinx develops when the normal flow of CSF fluid around the spinal cord or lower brain stem is disturbed. When syrinxes affect the brain stem, the condition is called syringobulbia.

What causes syringomyelia?

Syringomyelia may have several possible causes but most cases are associated with Chiari malformation—an abnormal condition in which brain tissue extends through the hole at the bottom of the skull (foramen magnum) and into the spinal canal, and obstructs the flow of CSF. Syringomyelia may also be caused by spinal cord injuries, spinal cord tumors, and damage caused by inflammation in around the spinal cord. In some cases, the cause is unknown (idiopathic).

What are the forms of syringomyelia?

There are two major forms of syringomyelia.

- **Congenital syringomyelia** (also known as communicating syringomyelia). In most cases, syringomyelia is caused by a Chiari malformation which may allow a syrinx to develop, most often in the spine's cervical (neck) region. Symptoms usually begin between the ages of 25 and 40. People with congenital syringomyelia may also have hydrocephalus, a buildup of excess CSF in the brain with enlargement of the cerebral ventricles. Straining or coughing can force CSF into the ventricles, causing the person to develop headache or even lose consciousness (so called cough syncope). In addition, they may have a disorder called arachnoiditis—an inflammation of the arachnoid, one of the three membranes that surrounds the spinal cord.

- **Acquired syringomyelia** (also known as primary spinal syringomyelia or noncommunicating syringomyelia). Causes of acquired syringomyelia include spinal cord injury, meningitis (an inflammation of brain and spinal cord membranes usually caused by an infection), arachnoiditis, tethered cord syndrome (a condition that is present at birth that causes the spinal cord to abnormally attach to the tissues in the lower spine, limiting its movement), a spinal cord tumor, and bleeding into the cord (hemorrhage).

What are symptoms of syringomyelia?

Symptoms of damage to the spinal cord vary among individuals according to where the syrinx forms, how large it is, and how long it extends. Symptoms develop slowly over time and may occur on one or both sides of the body. Sometimes coughing and straining can trigger symptoms, but they do not cause syringomyelia. Symptoms may include:

- pain
- progressive weakness in the arms and legs
- stiffness in the back, shoulders, neck, arms, or legs
- headaches
- loss of sensitivity to pain or hot and cold, especially in the hands
- numbness or tingling
- imbalance
- loss of bowel and bladder control

- problems with sexual function
- curvature of the spine (scoliosis) that may be the only symptom in children.

How is syringomyelia diagnosed?

To diagnose syringomyelia, a physician will review the person's medical history and perform a physical exam focusing on neurological function. The physician will also likely order imaging of the spine or brain. Sometimes, syringomyelia may be found during diagnostic imaging for another disorder.

Magnetic resonance imaging (MRI) is the most reliable way to diagnose syringomyelia. Computer-generated radio waves and a powerful magnetic field produce clear images of the brain and spinal cord. Using this test, a physician will be able to determine if there is a syrinx in the spine or another abnormality, such as a tumor.

In some cases a dynamic MRI may be performed to show the flow of fluid around the spinal cord and within the syrinx. Many images are taken in rapid succession to provide such dynamic views. A dye or contrast agent may be injected to enhance the MRI images.

How is syringomyelia treated?

The type of treatment for syringomyelia depends on the severity and progression of an individual's symptoms.

Monitoring

In the absence of symptoms, syringomyelia is usually not treated. In addition, a physician may recommend not treating the condition in individuals of advanced age or in cases where there is no progression of symptoms. However, people should be carefully monitored by a neurologist or neurosurgeon as symptoms can worsen over time. Individuals may also want to avoid activities that involve straining (e.g., lifting heavy objects, jumping) since these actions can trigger symptoms. People with an associated Chiari malformation are especially apt to experience headache with straining.

Surgery

Surgery is usually recommended for individuals with symptomatic or progressive syringomyelia. There are two general forms of treatment: restoration of normal CSF flow around the spinal cord, and direct drainage of the syrinx. The type of treatment depends on what is causing the symptoms.

- **Treating the Chiari malformation.**

The main goal of Chiari surgery is to provide more space at the base of the skull and upper neck. This reduces pressure on the brain and spinal cord and restores the normal flow of CSF. Surgery can allow the syrinx to drain, sometimes becoming smaller or even disappearing entirely. Symptoms may improve even if the syrinx remains the same size or is reduced only slightly.

Individuals should get treated sooner rather than later since delaying treatment can cause irreversible spinal cord damage. Syringomyelia can reoccur after surgery, making additional operations necessary.

- **Preventing a syrinx from forming or expanding after an injury.** In the case of trauma-related syringomyelia, the primary strategy is to prevent a syrinx from developing or growing in the first place. This somewhat controversial procedure (called expansive duraplasty) is done in a surgical operation on the spine that involves removing scar tissue around the spinal cord and sometimes adding a patch to expand the “dura,” the membrane that surrounds the spinal cord. By clearing and expanding the space around the spinal cord, the normal flow of CSF may be restored.
- **Removing the obstruction.** Surgical removal of obstructions such as scar tissue, bone from the spinal canal, or tumors can help restore the normal flow of CSF. If a tumor is causing syringomyelia, removing the tumor almost always eliminates the syrinx. Occasionally, radiation may also be used to shrink the tumor.
- **Draining the syrinx.** Especially if there is no associated Chiari malformation or tumor, it may be necessary to drain the syrinx. This is usually done in cases where the syrinx is growing or the cause of the CSF obstruction cannot be identified.

To perform this procedure a surgeon inserts a drain called a stent or shunt. A stent consists of a small tube that is inserted into the syrinx fluid and allows fluid from within the cord to drain into the space just outside the spinal cord. Similarly, a shunt consists of a flexible tube and a valve that drains the syrinx fluid into another part of the body, usually the abdomen. By draining the syrinx, both of these procedures can halt the progression of symptoms and relieve headache. However, these procedures carry the risk of spinal cord injury, infection, or bleeding, and may not help all individuals.

What research is being done?

The mission of the National Institute of Neurological Disorders and Stroke (NINDS) is to seek fundamental knowledge about the brain and nervous system and to use that knowledge to reduce the burden of neurological disease. The NINDS is a component of the National Institutes of Health (NIH), the leading supporter of biomedical research in the world.

Genetics and birth defects

Studies are underway to better understand the role of genetic factors in Chiari I malformation, the most common cause of syringomyelia. Individuals with a Chiari I malformation who also have a family member with either the abnormality or syringomyelia are being studied to identify the location of the gene(s) responsible for the malformation.

In some cases, birth defects may be associated with brain malformations that can cause syringomyelia. Learning how and when these defects occur during fetal development may help scientists develop strategies that can stop the formation of certain birth defects. The use of folic acid dietary supplements during pregnancy, for example, has been found to reduce the risk of birth defects of the brain and spinal cord, including malformations of the skull, brain, and spine.

Treatments

NINDS scientists are examining individuals who either have syringomyelia or are at risk of developing the disorder. They are investigating the factors that influence its development, progression, and response to treatment by recording more than 5 years of symptoms, muscle strength, overall function, and MRI findings from individuals who receive standard treatment for syringomyelia. Study results may allow scientists to provide more accurate recommendations to future patients with syringomyelia regarding optimal surgical or non-surgical treatments.

Also, NIH-funded scientists are trying to find ways to stop and reverse the cell damage caused by a spinal cord injury to help individuals recover their ability to move their muscles and other functions.

Diagnostic tools

Diagnostic technology is another area for continued research. Ongoing NIH-funded research is aimed at improving diagnostic imaging techniques to better visualize conditions in the spine, including syringomyelia, even before symptoms appear.

In addition, the NINDS is developing common data elements for Chiari Malformation. This will allow researchers to have a common scientific language that will improve data quality and opportunities to compare and combine data at multiple institutions.

Where can I get more information?

For more information on neurological disorders or research programs funded by the National Institute of Neurological Disorders and Stroke, contact the Institute's Brain Resources and Information Network (BRAIN) at:

BRAIN

P.O. Box 5801

Bethesda, MD 20824

800-352-9424

www.ninds.nih.gov

More information about research on syringomyelia supported by NINDS and other NIH Institutes and Centers can be found using NIH RePORTER (projectreporter.nih.gov), a searchable database of current and past research projects supported by NIH and other federal agencies. RePORTER also includes links to publications and resources from these projects.

Information also is available from the following organizations:

American Chronic Pain Association

P.O. Box 850
Rocklin, CA 95677-0850
916-632-0922
800-533-3231
www.theacpa.org

American Syringomyelia & Chiari Alliance Project

P.O. Box 1586
Longview, TX 75606-1586
903-236-7079
800- 272-7282
www.asap.org

Chiari and Syringomyelia Foundation

29 Crest Loop
Staten Island, NY 10312
718-966-2593
www.csfinfo.org

Christopher and Dana Reeve Foundation

636 Morris Turnpike, Suite 3A
Short Hills, NJ 07078
800-225-0292
www.christopherreeve.org

National Organization for Rare Disorders (NORD)

55 Kenosia Avenue
Danbury, CT 06810-1968
203-744-0100
Toll-free voicemail: 800-999- 6673
www.rarediseases.org

U.S. National Library of Medicine
National Institutes of Health/DHHS
8600 Rockville Pike
Bethesda, MD 20894
301-594-5983
888-346-3656
www.nlm.nih.gov



National Institute of
Neurological Disorders
and Stroke

NIH . . . Turning Discovery Into Health

Prepared by:
Office of Communications and Public Liaison
National Institute of Neurological
Disorders and Stroke
National Institutes of Health
Department of Health and Human Services
Bethesda, Maryland 20892-2540