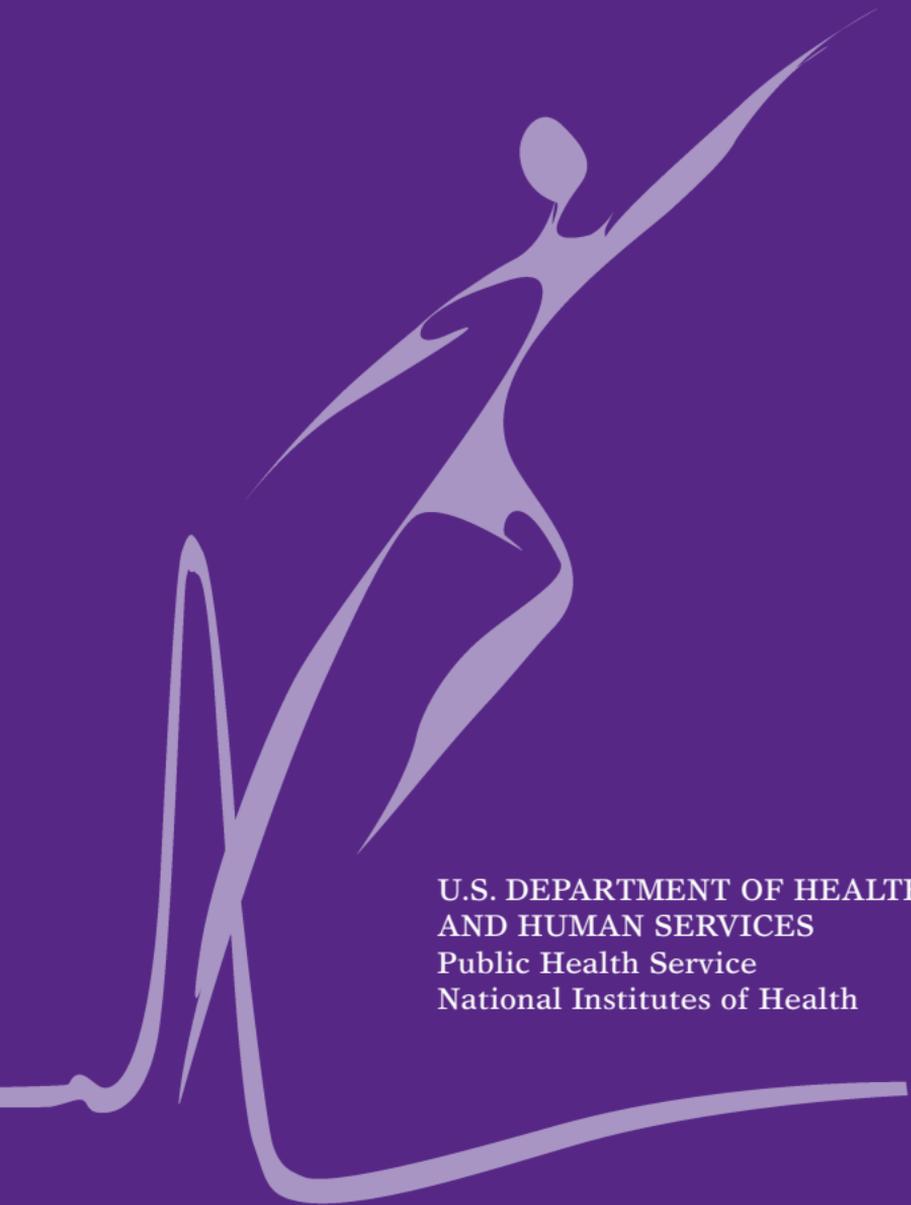


Syringomyelia

A stylized, light-colored graphic on a dark purple background. It features a silhouette of a person with their arms raised in a gesture of triumph or relief. Below the figure is a white waveform, similar to an ECG or EEG trace, with a prominent peak. The overall composition is minimalist and symbolic.

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

Syringomyelia

What is syringomyelia?

Syringomyelia (sear-IN-go-my-EEL-ya) is a disorder in which a cyst forms within the spinal cord. This cyst, called a syrinx, expands and elongates over time, destroying a portion of the spinal cord from its center and expanding outward. When a syrinx widens enough to affect nerve fibers that carry information from the brain to the extremities, this damage results in pain, weakness, and stiffness in the back, shoulders, arms, or legs. Other symptoms may include headaches and a loss of the ability to feel extremes of hot or cold, especially in the hands. Each patient experiences a different combination of symptoms depending on where in the spinal cord the syrinx forms and how far it expands.

Other more common disorders share the early symptoms of syringomyelia. In the past, this has made diagnosis difficult. But the widespread availability of an outpatient imaging procedure called magnetic resonance imaging (MRI) has significantly increased the number of syringomyelia cases diagnosed in the beginning stages of the disorder.

Because syringomyelia can occur in association with other conditions, estimates of the number of Americans with the disorder vary

widely, but a conservative estimate is that about 40,000 people in the United States are affected, with symptoms usually beginning in young adulthood. Signs of the disorder tend to develop slowly, although sudden onset may occur with coughing or straining. If not treated surgically, syringomyelia often leads to progressive weakness in the arms and legs, loss of hand sensation, loss of bladder and other functions, and chronic, severe pain.

What causes syringomyelia?

A watery, protective substance known as cerebrospinal fluid (CSF) normally flows around the spinal cord and brain, transporting nutrients and waste products. It also serves to cushion the brain. In early development, CSF also fills a small canal through the center of the spinal cord—the central canal—which then collapses normally over time.

A number of medical conditions can cause an obstruction in the normal flow of CSF, redirecting it into the central canal, and ultimately into the spinal cord itself. For reasons that are only now becoming clear, this redirected CSF fills the expanding central canal and results in syrinx formation. Pressure differences along the spine cause the fluid to move within the cyst. Physicians believe that it is this continual movement of fluid that builds pressure around and inside the spinal cord and results in cyst growth and further damage to the spinal cord tissue.

What are the different forms of syringomyelia?

Generally, there are two forms of syringomyelia. In most cases, the disorder is related to an abnormality of the brain called a Chiari I malformation, named after the physician who first characterized it. This anatomic abnormality causes the lower part of the cerebellum to protrude from its normal location in the back of the head into the cervical or neck portion of the spinal canal. A syrinx may then develop in the cervical region of the spinal cord. Because of the relationship that was once thought to exist between the brain and spinal cord in this type of syringomyelia, physicians sometimes refer to it as communicating syringomyelia. Symptoms usually begin between the ages of 25 and 40 and may worsen with straining or any activity that causes CSF pressure to fluctuate suddenly. Some patients, however, may have long periods of stability. Others with this form of the disorder also have hydrocephalus, in which CSF accumulates in the skull, or a condition called arachnoiditis, in which a covering of the spinal cord—the arachnoid membrane—is inflamed.

The second major form of syringomyelia occurs as a complication of trauma, meningitis, hemorrhage, a tumor, or arachnoiditis. Here, the syrinx or cyst develops in a segment of the spinal cord damaged by one of these conditions. The syrinx then starts to expand. This is sometimes referred to as noncommunicating syringomyelia. Symptoms may appear months or even years after the initial injury,

starting with pain, weakness, and sensory impairment originating at the site of trauma.

The primary symptom of post-traumatic syringomyelia is pain, which may spread upward from the site of injury. Symptoms, such as pain, numbness, weakness, and disruption in temperature sensation, may occur on one or both sides of the body. Syringomyelia can also adversely affect sweating, sexual function, and, later, bladder and bowel control.

In addition, one form of the disorder involves a part of the brain called the brainstem. The brainstem controls many of our vital functions, such as respiration and heartbeat. When syrinxes affect the brainstem, the condition is called syringobulbia. Some cases of syringomyelia are familial, although this is rare.

How is syringomyelia diagnosed?

Physicians now use magnetic resonance imaging (MRI) to diagnose syringomyelia. The MR imager takes pictures of body structures, such as the brain and spinal cord, in vivid detail. This test will show the syrinx in the spine as well as other conditions, such as the presence of a tumor. MRI is safe, painless, and informative and has greatly improved the diagnosis of syringomyelia. Images taken in rapid succession can be used for “dynamic imaging” (in “*cine mode*”) to observe the fluid flowing around the spinal cord and within the syrinx.

The physician may order additional tests to help confirm the diagnosis. One of these is called electromyography (EMG), which measures muscle weakness. The doctor may also

wish to test CSF pressure levels and to analyze the CSF by performing a lumbar puncture. In addition, computed tomography (CT) scans of a patient's head may reveal the presence of tumors and other abnormalities such as hydrocephalus.

Like MRI and CT scans, another test, called a myelogram, takes x-ray-like pictures and requires a contrast medium or dye to do so. Since the introduction of MRI this test is rarely necessary to diagnose syringomyelia.

How is syringomyelia treated?

Surgery is usually recommended for syringomyelia patients. The main goal of surgery is to provide more space for the cerebellum (Chiari malformation) at the base of the skull and upper neck, without entering the brain or spinal cord. This results in flattening or disappearance of the primary cavity. If a tumor is causing syringomyelia, removal of the tumor is the treatment of choice and almost always eliminates the syrinx.

Surgery results in stabilization or modest improvement in symptoms for most patients. Delay in treatment may result in irreversible spinal cord injury. Recurrence of syringomyelia after surgery may make additional operations necessary; these may not be completely successful over the long term.

In some patients it may be necessary to drain the syrinx, which can be accomplished using a catheter, drainage tubes, and valves. This system is also known as a shunt. Shunts are used in both the communicating and noncommunicating forms of the disorder.

First, the surgeon must locate the syrinx. Then, the shunt is placed into it with the other end draining the syrinx fluid into a cavity, usually the abdomen. This type of shunt is called a syringoperitoneal shunt. A shunt of CSF from the brain to the abdomen is called a ventriculoperitoneal shunt and is used in cases involving hydrocephalus. By draining syrinx fluid or CSF, a shunt can arrest the progression of symptoms and relieve pain, headache, and tightness. Without correction, symptoms generally continue.

The decision to use a shunt requires extensive discussion between doctor and patient, as this procedure carries with it the risk of injury to the spinal cord, infection, blockage, or hemorrhage and may not necessarily work for all patients.

In the case of trauma-related syringomyelia, the surgeon operates at the level of the initial injury. Until the 1990s, the most common approach was to collapse the cyst in surgery and insert a tube or shunt to prevent its re-expansion. Because shunts routinely become clogged and require multiple operations, many surgeons now consider this option only as a last resort. Instead, surgeons now expand the space around the spinal cord by realigning the vertebrae or discs that are narrowing the spinal column. They then add a patch to expand the “dura,” the membrane that surrounds the spinal cord and contains the CSF (a procedure called “expansive duraplasty”). It is also considered

important to remove scar tissue attached to the membranes that “tether” the cord in place and prevent the free flow of CSF around it.

Drugs have no curative value as a treatment for syringomyelia. Radiation is used rarely and is of little benefit except in the presence of a tumor. In these cases, it can halt the extension of a cavity and may help to alleviate pain.

In the absence of symptoms, syringomyelia is usually not treated. In addition, a physician may recommend not treating the condition in patients of advanced age or in cases where there is no progression of symptoms. Whether treated or not, many patients will be told to avoid activities that involve straining.

Many spinal cord injured individuals have a cyst at the site of their original injury. These cysts do not always require treatment, although if they grow larger or begin to cause symptoms treatment may be warranted.

What research is being done?

The precise causes of syringomyelia are still unknown. Scientists at the National Institute of Neurological Disorders and Stroke (NINDS) in Bethesda, Maryland, and at grantee institutions across the country continue to explore the mechanisms that lead to the formation of syrinxes in the spinal cord. For instance, Institute investigators have found that as the heart beats, the syrinx fluid is abruptly forced downward.

They have also demonstrated the presence of a block to the free flow of CSF that normally occurs in and out of the head during each heartbeat. In the past decade, clinicians and researchers used this new understanding to improve the standard of care for patients experiencing loss of function due to expanding syrinxes. They are now studying how syrinxes first form in disorders known to produce syringomyelia. In some cases, chronic enlargement of the cord and other changes might be detected earlier than is currently the case, allowing surgical treatment before loss of function becomes permanent.

NINDS scientist-physicians are conducting clinical studies at the NIH to learn more about the mechanisms of syringomyelia, for example, how CSF pressure and flow contribute to the progression of the disorder. In these studies patients with progressive syringomyelia undergo clinically indicated tests and research tests as well as standard surgical care for the disorder.

In the 1990s, two early-phase clinical trials evaluated the safety of using embryonic tissue grafts to seal syrinxes in a few patients. Cells were grafted, the cyst was drained, and the space in the spinal column surrounding the spinal cord was expanded. These pilot studies indicated that the grafts were safe and the cysts did not re-expand in areas where surviving graft tissue was detectable on MR images. Researchers are actively

studying cell grafting into damaged areas of the spinal cord in hopes of preventing progressive degeneration and restoring function. The idea of using cell grafts to obliterate spinal cysts permanently may be explored in future studies, as optimal cell grafting techniques are developed.

It is also important to understand the role of birth defects in the development of hindbrain malformations that can lead to syringomyelia. Learning when these defects occur during the development of the fetus can help us understand this and similar disorders and may lead to preventive treatment that can stop the formation of many birth abnormalities. Dietary supplements of folic acid during pregnancy have already been found to reduce the number of cases of certain birth defects.

Diagnostic technology is another area for continued research. Already, MRI has enabled scientists to see conditions in the spine, including syringomyelia, even before symptoms appear. A new technology known as dynamic MRI allows investigators to view spinal fluid pulsating within the syrinx. CT scans allow physicians to see abnormalities in the brain, and other diagnostic tests have also improved greatly with the availability of new, non-toxic, contrast dyes. Patients can expect even better techniques to become available in the future from the research efforts of scientists today.

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, Maryland 20824
301-496-5751
800-352-9424
www.ninds.nih.gov

Information also is available from the following organizations:

American Chronic Pain Association (ACPA)

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

American Syringomyelia Alliance Project (ASAP)

P.O. Box 1586
Longview, Texas 75606-1586
903-236-7079
800-ASAP-282 (272-7282)
www.asap.org

**Christopher Reeve Foundation and
Resource Center**

636 Morris Turnpike
Suite 3A
Short Hills, New Jersey 07078
973-379-2690
800-225-0292
www.christopherreeve.org

March of Dimes Birth Defects Foundation

1275 Mamaroneck Avenue
White Plains, New York 10605
914-428-7100
888-MODIMES (663-4637)
www.marchofdimes.com

**National Chronic Pain Outreach
Association (NCPOA)**

P.O. Box 274
Millboro, Virginia 24460
540-862-9437
www.chronicpain.org

**National Organization for Rare Disorders
(NORD)**

P.O. Box 1968
(55 Kenosia Avenue)
Danbury, Connecticut 06813-1968
203-744-0100
Voice Mail: 800-999-NORD (6673)
www.rarediseases.org

National Spinal Cord Injury Association

6701 Democracy Boulevard
Suite 300
Bethesda, Maryland 20817
301-214-4006
800-962-9629
www.spinalcord.org

Paralyzed Veterans of America (PVA)

801 18th Street, NW

Washington, DC 20006-3517

202-USA-1300 (872-1300)

800-424-8200

www.pva.org

Spina Bifida Association of America

4590 MacArthur Boulevard, NW

Suite 250

Washington, DC 20007-4266

202-944-3285

800-621-3141

www.sbaa.org

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Fergus Falls, Minnesota 56537

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