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Syringomyelia Fact Sheet

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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. As a syrinx widens it compresses and injures nerve fibers that carry information from the brain to the extremities. Damage to the spinal cord often leads to progressive weakness in the arms and legs, stiffness in the back, shoulders, arms, or legs, and chronic, severe pain. Other symptoms may include headaches, a loss of the ability to feel extremes of hot or cold (especially in the hands), and loss of bladder and other functions. Each individual experiences a different combination of symptoms depending on where in the spinal cord the syrinx forms and how far it extends. Signs of the disorder tend to develop slowly, although sudden onset may occur with coughing or straining.

Many disorders share the early symptoms of syringomyelia, which also can occur in association with other conditions. Estimates of the number of Americans with syringomyelia 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. Some cases of syringomyelia occur in more than one family member, although this is rare.

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 acts as a cushion to protect the brain and spinal cord. In early development, CSF also fills a small canal that runs 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 excess fluid into the spinal cord and central canal. This redirected CSF accumulates within the tissue of the spinal cord and forms a fluid-filled syrinx. Pressure differences along the spine cause the fluid to move within the syrinx and outside the spinal cord. Physicians believe that this continual movement of fluid in and around the spinal cord 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 one of the physicians 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 individuals, however, may have long periods of functional stability. Some people with this form of the disorder also have hydrocephalus, in which CSF accumulates in the brain, 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.

Syringomyelia may also involve 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.

How is syringomyelia diagnosed?

Diagnostic imaging has significantly increased the number of syringomyelia cases detected in the beginning stages of the disorder. Physicians primarily use magnetic resonance imaging (MRI) to diagnose syringomyelia. Computer-generated radio waves and a powerful magnetic field produce images 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. 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. Computed tomography (CT), which uses x-rays and a computerized scanner to produce cross-sectional images of the body or an organ structure, may reveal the presence of tumors and other abnormalities such as hydrocephalus. 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 individuals with syringomyelia. The type of surgery and its location depend on the type of syringomyelia. In Chiari malformation, the main goal of surgery is to provide more space at the base of the skull and upper neck, without entering the brain or spinal cord. This results in the primary cyst becoming much smaller. Surgery results in stabilization or modest improvement in symptoms for most individuals with Chiari malformation. 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 individuals it may be necessary to drain the syrinx, which can be accomplished using a catheter, drainage tubes, and valves. This system is 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 halt 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 the surgeon and the individual, 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 people.

If a tumor is causing syringomyelia, removing the tumor is the treatment of choice and almost always eliminates the syrinx.

In the case of trauma-related syringomyelia, the preferred surgical approach is to operate at the level of the initial injury to expand the space around the spinal cord and decrease fluid volume. This operation is performed outside the spinal cord. An alternate operation is to place a shunt in the syrinx, which requires a hole to be made in the spinal cord. Shunts may injure the spinal cord and may require replacement if they clog over time. Many surgeons now consider shunt placement only as a last resort. Instead, surgeons usually choose to expand the space around the spinal cord. This is done by removing scar tissue that “tethers” the cord in place and prevents the free flow of CSF around it, and adding a patch to expand the “dura,” the membrane that surrounds the spinal cord and contains the CSF (a procedure called expansive duraplasty). In some cases, the vertebrae may need to be realigned to correct spinal deformity that is narrowing the spinal column.

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

Drugs have no curative value as a treatment for syringomyelia but may be used to ease pain. 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 individuals of advanced age or in cases where there is no progression of symptoms. Whether treated or not, many individuals will be told to avoid activities that involve straining.

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 people who experience 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 or tethering of the spinal 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 abnormal CSF flow contributes to the progression of the disorder. In these studies individuals with progressive syringomyelia undergo clinical procedures and research tests as well as standard surgical care for the disorder.

Studies are under way to better understand the genetic factors related to a Chiari I malformation. Individuals with a Chiari I malformation who also have a family member with either the abnormality or syringomyelia are being studied to discover the location of the gene(s) responsible for the malformation.

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 been found to reduce the number of cases of certain birth defects.

Diagnostic technology is another area for continued research. 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. Other diagnostic tests have also improved greatly with the availability of new, non-toxic, contrast dyes. Improved techniques are expected 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, MD 20824
(800) 352-9424
http://www.ninds.nih.gov

Information also is available from the following organizations:

American Chronic Pain Association (ACPA)
P.O. Box 850
Rocklin, CA 95677-0850
ACPA@theacpa.org
http://www.theacpa.org
Tel: 916-632-0922 800-533-3231
Fax: 916-652-8190

American Syringomyelia & Chiari Alliance Project (ASAP)
P.O. Box 1586
Longview, TX 75606-1586
info@asap.org
http://www.asap.org
Tel: 903-236-7079 800-ASAP-282 (272-7282)
Fax: 903-757-7456

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Suite 3A
Short Hills, NJ 07078
informations@christopherreeve.org
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Tel: 973-379-2690 800-225-0292
Fax: 973-912-9433

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White Plains, NY 10605
askus@marchofdimes.com
http://www.marchofdimes.com
Tel: 914-997-4488 888-MODIMES (663-4637)
Fax: 914-428-8203

National Organization for Rare Disorders (NORD)
55 Kenosia Avenue
Danbury, CT 06810
orphan@rarediseases.org
http://www.rarediseases.org
Tel: 203-744-0100 Voice Mail 800-999-NORD (6673)
Fax: 203-798-2291

National Spinal Cord Injury Association
75-20 Astoria Blvd
Suite 120
East Elmhurst, NY 11370-1177
info@spinalcord.org
http://www.spinalcord.org
Tel: 800-962-9629
Fax: 866-387-2196

Paralyzed Veterans of America (PVA)
801 18th Street, NW
Washington, DC 20006-3517
info@pva.org
http://www.pva.org
Tel: 202-USA-1300 (872-1300) 800-555-9140
Fax: 202-785-4452

Spina Bifida Association
4590 MacArthur Blvd. NW
Suite 250
Washington, DC 20007-4266
sbaa@sbaa.org
http://www.spinabifidaassociation.org
Tel: 202-944-3285 800-621-3141
Fax: 202-944-3295

Spinal Cord Society
19051 County Highway 1
Fergus Falls, MN 56537
scs-nc@nc.rr.com
http://scsus.org/
Tel: 218-739-5252 or 218-739-5261
Fax: 218-739-5262

Chiari & Syringomyelia Foundation
29 Crest Loop
Staten Island, NY 10312
info@CSFinfo.org
http://www.csfinfo.org
Tel: 718-966-2593
Fax: 718-966-2593 (Call First)

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Publicaciones en Español

- **Siringomielia**

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