Hydromyelia

Definition

Hydromyelia (HM) is a condition characterized by widening of the central canal of the spinal cord. Fluid can accumulate in this space, creating increased pressure on the spinal cord. The term hydromyelia is sometimes used interchangeably with a closely related condition, syringomyelia (or syringohydromyelia). Syringomyelia (SM) is a condition in which fluid collects in the area of the spinal cord that is outside the central canal. The end result of hydromyelia and syringomyelia is essentially the same: an abnormal cyst (collection of fluid) in the spinal cord that is associated with a wide range of neurological complaints and signs. For simplicity, the term syringomyelia is used to refer to a fluid−filled cyst in the spinal cord that is inside or outside of the central canal.

Description

Syringomyelia is a variable condition in which the symptoms depend on the location and extent of the cavitation (hollowing out) of the cord. Over time, the expansion and elongation of the fluid−filled cavity (or cyst) can destroy the center of the spinal cord. This damage to the spinal cord results in pain, weakness, and loss of sensation for the affected individual. Syringomyelia may be an isolated finding or may be found in association with a syndrome that disrupts the flow of cerebral spinal fluid (CSF), such as the Arnold−Chiari malformation or the Dandy−Walker malformation.

The earliest known description of cystic dilatation (widening) of the spinal cord dates back to the sixteenth century. The terms syringomyelia and hydromyelia were first used in published reports in 1827 and 1859, respectively.

Demographics

Syringomyelia occurs across all races and ethnic groups and affects both children and adults. Although syringomyelia usually appears in midlife, it can occur at any age. Estimates of the incidence of syringomyelia

vary and range from 1 in 18,000 to as high as 1 in 1,300 people in the United States.

Causes and symptoms

The causes of syringomyelia are not well understood. It is thought that syringomyelia occurs when one or more factors interfere with the normal development of the spinal canal during formation of the embryo or when factors such as trauma to the spinal cord, infection, or a mass (such as tumor) interfere with the fluid dynamics in the spinal cord. Arnold−Chiari malformation is the leading cause of syringomyelia. Syringomyelia occurs in as many as onequarter of people who have a spinal cord injury. Various theories have been postulated to explain how movement of cerebrospinal fluid and pressure in the central nervous system (the brain and spinal cord) interact to produce this defect. In some cases, genetic factors may play a role in the development of this condition.

The symptoms of syringomyelia can be quite variable and depend upon the location and extent of the cyst. Common symptoms of syringomyelia in affected individuals include:

- extreme pain or "heavy" feeling in the neck; shoulders are usually numb
- headaches
- leg or hand weakness
- numbness or loss of sensation in the hands and feet
- problems with walking
- loss of bowel and bladder control
- spasticity and paralysis of the legs
- visual disturbances
- ataxia

- speech problems
- scoliosis (curvature of the spine)

Diagnosis

Diagnosis of syringomyelia is based on neurological exam and results of neuroradiological imaging studies. The neurological exam of an affected individual will show loss of sensation in the hands, balance problems, decreased strength, difficulty walking or an abnormal gait, and abnormal reflexes. Some people with no symptoms or mild symptoms are diagnosed with syringomyelia incidentally in the course of evaluation for another condition. Imaging studies used to diagnose syringomyelia include magnetic resonance imaging (MRI), CINE MRI (a type of MRI that shows the flow of cerebrospinal fluid), and electromyography (EMG). In some cases it may be technically difficult to distinguish hydromyelia from syringomyelia.

Treatment

Currently, there is no cure for syringomyelia. Neuro−surgery is the primary method of treatment for this condition. Surgery tends to be reserved for those individuals with moderate or severe neurological problems. The goal of the various neurosurgical techniques is to restore normal flow of cerebral spinal fluid. There are four main categories of procedures:

- decompression procedures
- laminectomy and syringostomy
- terminal ventriculostomy
- percutaneous aspiration

Surgical interventions for syringomyelia have limitations and carry risks for potentially severe complications. The decision to operate is generally based on the severity of symptoms and the findings on MRI or other imaging studies. Patients are followed closely after surgery for signs of further neurological impairment. Some patients will need to undergo more than one surgery.

Treatment team

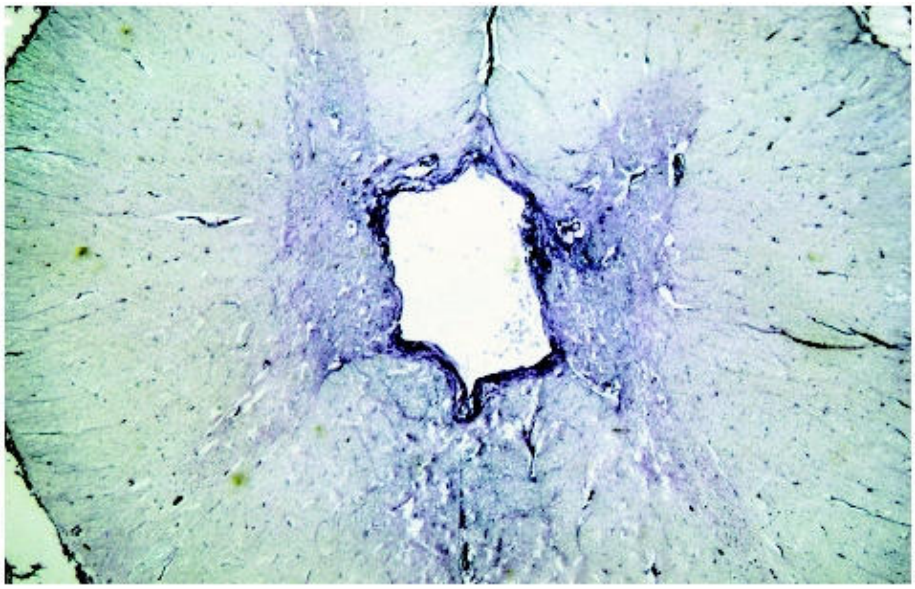
Management of syringomyelia requires a multidisciplinary approach. In addition to the patient's primary health care professionals, medical professionals involved in the care of patients with syringomyelia generally include a neurologist and a neurosurgeon. Additional specialists in pain management and rehabilitation may also be needed.

Recovery and rehabilitation

Patients with syringomyelia may require a wide range of rehabilitation services including physical therapy, occupational therapy, and speech therapy to help them compensate for weakness and loss of function. Chronic pain can pose a significant problem for some patients. Management of chronic pain may include prescription and non−prescription medications, physical therapy, occupational therapy, medical procedures such as nerve blocks or trigger point injections, psychological therapy, and chiropractics.

Clinical trials

As of early 2004, there were three clinical trials for patients with syringomyelia, all of which are sponsored by the National Institute of Neurological Disorders and Stroke (NINDS). There is a study (Study and Surgical Treatment of Syringomyelia) to establish the mechanism(s) of progression of primarily spinal syringomyelia (PSS). More information on this study can be obtained at http://clinicalstudies.info.nih.gov/detail/A_2001−N−0085.html or by contacting the patient recruitment and public liaison office at (800) 411−1222 or prpl@mail.cc.nih.gov. In another study (Establishing the Physiology of Syringomyelia), researchers would like to learn more about how the CSF pressure and flow contribute to the progression of syringomyelia. More information can be obtained at http://clinicalstudies.info.nih.gov/detail/A_1992−N−0226.html or by contacting the patient recruitment and public liaison office. Finally, there is a study (Genetic Analysis of the Chiari I Malformation) whose purpose is to better understand the genetic factors related to the Chiari I malformation. More information can be found at http://clinicalstudies.info.nih.gov/detail/A_2000−N−0089.html or by contacting the patient recruitment and public liaison office. There is also an ongoing genetic research study for Chiari type I malformation and syringomyelia (CMI/S) to determine whether or not there is a genetic component to CMI/S. Interested patients and families may find more information at the Center for Human Genetics at Duke University at http://wwwchg.mc.duke.edu/patients/cms.html or by contacting the center at (800) 283−4316 or syringo@dnadoc.mc.duke.edu.



The widened spinal cord canal associated with hydromyelia. (Custom Medical Stock Photo. *All Rights Reserved.*)

Prognosis

The course of syringomyelia is not well defined. Some untreated patients will experience a spontaneous remission of symptoms. Among treated patients, some will have a permanent end to their neurological deficits whereas others will only experience temporary relief of symptoms. Long−term studies of affected patients are needed to better understand the natural history and prognosis of this condition.

Resources

BOOKS

- Graham, D. I., and P. L. Lantos, eds. *Greenfield's Neuropathology*, volume I, 7th edition. London: Arnold, 2002.
- Parker, James N., MD, and Philip M. Parker, PhD, eds. *The Official Parent's Sourcebook on Syringomyelia: A Revised and Updated Directory for the Internet Age.* San Diego, CA: ICON Health Publications, 2002.
- Klekamp, Joerg, and Madjid Samii. *Syringomyelia: Diagnosis and Treatment*, 1st edition. New York: Springer Verlag, 2001.

PERIODICALS

- Caldarelli, M., and C. Di Rocco. "Diagnosis of Chiari I Malformation and Related Syringomyelia: Radiological and Neurophysiological Studies." *Child's Nervous System* 53 (March 2004): epublication, ahead of print.
- Kyoshima K., and E. I. Bogdanov. "Spontaneous Resolution of Syringomyelia: Report of Two Cases and Review of the Literature." *Neurosurgery* 53 (Sept 2003): 762–9.
- Wisoff, J. H. "Hydromyelia: A Critical Review." *Child's Nervous System* 4 (1988): 1–8.

WEBSITES

- *American Syringomyelia Alliance Project, Inc. Home Page.* (June 2, 2004). <http://www.asap.org.html>.
- *Chiari and Syringomyelia News Home Page.* (June 2, 2004). <http://www.chiari−syringo−news.com/.html>.
- The National Institute of Neurological Disorders and Stroke (NINDS). *Hydromyelia Information Page*. (June 2, 2004). <http://www.ninds.nih.gov/health_and_medical/disorders/hydr... .
- The National Institute of Neurological Disorders and Stroke (NINDS). *Syringomyelia Information Page*. (June 2, 2004). <http://www.ninds.nih.gov/health_and_medical/disorders/syri... .

ORGANIZATIONS

- American Syringomyelia Alliance Project, Inc. P. O. Box 1586, Longview, TX 75606−1586. (903) 236−7079 or (800) ASAP−282; Fax: (903) 757−7456. info@ASAP.org. <http://www.asap.org>.
- American Chronic Pain Association (ACPA). P.O. Box 850, Rocklin, CA 95677−0850. (916) 632−0922 or (800) 533−3231; Fax: (916) 632−3208. ACPA@pacbell.net. <http://www.theacpa.org>.
- National Spinal Cord Injury Association. 6701 Democracy Blvd. #300−9, Bethesda, MD 20817. (301) 214−4006 or (800) 962−9629; Fax: (301) 881−9817. info@spinalcord.org. <http://www.spinalcord.org>.

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