Key Points

1. Dr. Ulrich Batzdorf is a widely regarded expert on Chiari and syringomyelia
2. Published a review on primary syringomyelia, or syringomyelia not associated with Chiari
3. Primary syringomyelia still involves disruption of the flow of CSF, but lower in the spine
4. Can be due to scarring from trauma, scarring from inflammation, arachnoid blockage, or tumors
5. Surgical goal is to remove the blockage and restore normal CSF flow
6. Outcomes are not as good as Chiari related surgery; out of 64 patients, only 15 improved
7. Shunting the syrinx directly should be used when there are no other choices
8. Patients often need treatment for spasticity and neuropathic pain after surgery

Definitions

arachnoid veil - like the dura, the arachnoid is one of the layers of the covering of the brain and spine, sometimes it can attach to other surfaces in a veil like pattern and disrupt CSF flow

central canal - tube like center of the spinal cord; the central canal tends to collapse as people age

cine MRI - type of MRI which can show motion, such as the flow of spinal fluid

craniovertebral junction - region where the skull meets the spine

CT - computed tomography; imaging technique which uses X-rays taken at many angles to construct images of internal body structures

duraplasty - surgical technique which involves sewing a patch into the dura (the covering of the brain and spine)

Primary Syringomyelia Difficult To Treat

January 20, 2006 -- A research publication (can't remember which one) once said that syringomyelia is not really an entity by itself, because it is always secondary to something else. The vast majority of syringomyelia cases are secondary to problems in the craniovertebral region, such as Chiari; however syringomyelia can be due to other causes such as trauma, infection, and tumors.

Syringomyelia not associated with Chiari is, somewhat confusingly, referred to as primary syringomyelia, and was the focus of a recent publication by one of the most highly regarded experts on Chiari and syringomyelia. Dr. Ulrich Batzdorf reported on his experiences with 64 primary syringomyelia patients in the December, 2005 issue of the Journal of Neurosurgery: Spine.

Much like Chiari related syringomyelia, Dr. Batzdorf reports that primary syringomyelia involves disruption of the natural flow of cerebrospinal fluid. In Chiari, this disruption occurs at the craniovertebral junction, in primary syringomyelia, the disruption occurs elsewhere in the spine and can be due to injury, infection, surgery, tumors, and arachnoid webs (see Table 1).

Figure 1
Surgically Treated Primary Syringomyelia By Cause, Plus Number of Surgeries (64 Patients)

Cause	# of Cases	# of Surgeries
traumatic scar/bone	26	58
post inflammation	16	23
tumor	11	12
arachnoid cyst	4	5
congenital tethering	2	2
residual central canal	1	1
unknown	4	4

Of the multiple causes of primary syringomyelia, post-traumatic SM is probably the most widely studied and reported on. After a traumatic spinal injury, bone fragments and scar tissue can lead to disruption of the CSF flow, resulting in the formation of a syrinx months to years after the initial injury. Estimates on how common post-traumatic SM is vary widely and range from as few as 2% to as many as 64% of spinal injury patients.

Post-inflammatory syringomyelia occurs when scarring develops after an inflammatory response due to an infection, such as meningitis. Inflammation can also occur in some people as a reaction to the introduction of a chemical, such as a contrast agent which was used many years ago. Unlike a localized scar from an injury, inflammation from meningitis can lead to widespread scarring along the length of the spine and result in more extensive syrinxes.

Prior surgery can also result in scarring which in turn causes syringomyelia. Procedure which open the dura to remove abnormal growths, even if the growths are benign, can lead to problems years later. It is not known whether unrecognized bleeding can cause syringomyelia, or if some people are just more prone to developing scar tissue than others.

As with Chiari related SM, the exact mechanisms involved in primary syrinx formation are not well understood. It is believed however, and there is evidence to support, that due to an obstruction, CSF enters the spinal tissue through the perivascular spaces. Perivascular spaces are small gaps between the outside wall of veins and arteries and the nearby tissue which they feed. It is believed that CSF flows through these spaces along the arteries and begins to collect in the spinal tissue itself.

Diagnosing primary syringomyelia is a little different than Chiari related SM. While an MRI can identify a syrinx, it does not always present a good picture of the underlying obstruction which led to the syrinx forming in the first place. Dr. Batzdorf strongly recommends using myelography, which involves injecting a contrast dye and a CT scan, to obtain a better understanding of the situation prior to surgery. Interestingly, cine-MRI, which is becoming standard in evaluating Chiari, is not as widely used, nor as successful in evaluating primary syringomyelia.

Because primary syringomyelia is thought to be due to obstructions to CSF flow, the first line of treatment is to surgically remove the obstruction and restore the natural flow of CSF. Only if this doesn't work, according to Dr. Batzdorf, should shunting be considered.

foramen magnum - opening at the base of the skull through which the brain connects to the spinal cord

laminectomy - surgical technique which involves removing part of one or more vertebra

myelography - imaging technique which combines a contrast agent injected into the spinal fluid and X-rays

neuropathic pain - pain due to nerve damage; can be very difficult to treat

perivascular spaces - small spaces on the outside of veins and arteries as they penetrate the spinal tissue

primary syringomyelia - syringomyelia which is not due to a craniovertebral abnormality, such as Chiari

spasticity - abnormal stiffness of muscles

cerebellar tonsils - portion of the cerebellum located at the bottom, so named because of their shape

cerebrospinal fluid (CSF) - clear liquid in the brain and spinal cord, acts as a shock absorber

Chiari malformation I - condition where the cerebellar tonsils are displaced out of the skull area into the spinal area, causing compression of brain tissue and disruption of CSF flow

MRI - magnetic resonance imaging; large device which uses strong magnetic fields to produce images of soft tissue inside the human body

syringomyelia (SM) - neurological condition where a fluid filled cyst forms in the spinal cord

syrinx - fluid filled cyst in the spinal cord

tonsillar herniation - descent of the cerebellar tonsils into the spinal area; often measured in mm

Source

Source: : Batzdorf U. Primary spinal syringomyelia. *Invited submission from the joint section meeting on disorders of the spine and peripheral nerves,* March 2005. J Neurosurg Spine. 2005 Dec;3(6):429-35.

Unfortunately, the results from surgery for primary syringomyelia are generally not as good as what is seen with a Chiari decompression (see Table 2). Out of 64 patients Batzdorf operated on, only 15 improved, while another 18 stabilized. This means that only about half of the patients had what could be considered a successful outcome. Additionally, 22 out of the 64 patients, nearly 1/3, required more than one surgery. Batzdorf does point out however, that in his experience, the outcomes for patients with localized problems - such as from tumors or small scars - tends to be better than when the problem is more diffuse, as in cases of post-inflammatory syringomyelia.

Figure 2
Surgical Outcome (64 Patients)

Outcome	# of Patients
Improved	15
Stabilized	18
Worsened	8
Required Reoperation	22
No Information	1

As with their Chiari brethren, the problems of primary syringomyelia patients often don't end with surgery. Spasticity, which is unusual stiffness of muscles, and neuropathic pain are all too common and need to be addressed if they are causing problems.

Primary syringomyelia is much less common than Chiari and likely less understood as well. There is very little published on the subject and in many cases treatment is difficult at best.

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