Key Points

1. In a small number of people, there is no identifiable cause of syringomyelia (SM), this is known as idiopathic SM
2. Recently, some doctors have begun treating idiopathic SM using standard Chiari decompression surgery with success
3. This study examined the clinical and MRI characteristics of a group of idiopathic SM patients, a group of Chiari SM patients, and a group of normal subjects
4. The clinical symptoms between the idiopathic and Chiari groups were the same
5. Both the idiopathic and Chiari groups had small posterior fossas and narrow CSF pathways relative to the normal subjects
6. The syrinxes of the Chiari group were wider than the idiopathic group
7. Study provides more evidence that Chiari surgery may benefit people with idiopathic SM

Definitions

ataxia - loss of coordination and balance due to nerve damage; trouble walking

atrophy - wasting away, withering

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

dorsal - referring to the back of something, can also refer to the top

duraplasty - surgical technique where a patch is sewn into the dura, the tough covering of the brain and spinal cord

etiology - the cause of a disease

foramen magnum - large opening

Taking The Unknown Out Of Idiopathic Syringomyelia

There are a lot of unknowns about syringomyelia, and by definition idiopathic SM is one of them. While the vast majority of SM cases are due to Chiari malformations, tumors, or trauma, for some people, there is no readily identifiable cause of their condition. These cases are referred to as idiopathic, meaning of unknown origin.

Without a tumor to remove, or a Chiari to decompress, for a long time doctors were unsure how to treat idiopathic SM. Recently however, some surgeons have shown - on a small number of cases - that despite the lack of a Chiari malformation, standard Chiari decompression surgery can help some cases of idiopathic SM. The term Chiari 0 has even been used to describe such cases. In addition, a large study (by Milhorat) showed that some people have syringomyelia with malformations less than 5mm - the classic definition of Chiari.

Building upon this evidence, Dr. Bogdanov, from the Kazan State Medical University in Russia, in collaboration with scientists from Russia, the NIH in the US, and the University of Saarland in Germany, theorized that if the same surgery is useful in treating Chiari related SM and idiopathic SM, then the underlying cause of the syrinx was probably the same in both cases. Specifically, they believe that the theory that Chiari develops as a result of a small posterior fossa - which does not have enough room for a normally developed brain - extends to cases of idiopathic SM.

To prove their theory, Bogdanov and his colleagues compared the clinical symptoms and MRI characteristics of 17 people with idiopathic SM, 17 people with Chiari related SM, and 32 normal people. All subjects underwent a clinical exam and a brain and spinal MRI. The MRI's were then reviewed and 11 distinct measurements were made in and around the head-spine junction. Measurements included - but were not limited to - the height of the posterior fossa, an angle which showed how curved the base of the skull was, the width of the CSF pathways in front of and behind the spinal cord, the length of the tonsillar herniation, and the width of the syrinx. The group published their results in the March, 2004 issue of the journal Neurology.

The scientists found that there was no real difference in clinical symptoms between the idiopathic and Chiari groups (see Fig 1). In addition, both SM groups had measurements which indicated smaller posterior fossas, flatter skull bases, and narrower CSF spaces than the normal subjects. The evidence supported their theory that idiopathic SM patients have smaller posterior fossas just like patients with Chiari related SM. The authors believe that in these cases, even if the cerebellum has not descended, with the narrow CSF pathways, the normal flow of CSF is disrupted and can lead to a syrinx forming.

While the two syringomyelia groups were similar when compared to the normal group, there were also some interesting differences between them. As might be expected the CSF pathway around the cerebellar tonsils was much smaller in the Chiari cases than in the idiopathic cases and in addition the syrinxes were larger in the Chiari cases. The researchers found it noteworthy that even though the Chiari group had wider syrinxes, the clinical symptoms - and presumably the damage from the syrinxes - were essentially the same.

The authors believe one explanation is that the MRI's for the idiopathic subjects were taken after the syrinxes had peaked in size. It is pretty well accepted in the medical community that given enough time, syrinxes will peak in size, but overtime will find ways to drain and become somewhat smaller. In addition, neurological damage can be rapid when a syrinx first forms but the progress tends to slow down over time and the symptoms become unrelated to the size of the syrinx. In fact, several studies have found very little correlation between syrinx size and symptoms.

Despite the minor differences between the idiopathic and Chiari groups, the evidence is mounting that the underlying mechanism for many idiopathic syringomyelia cases is similar to Chiari related SM. The obvious implication of this is that the same treatment used for Chiari related SM - namely decompression surgery - may work for idiopathic syringomyelia cases as well.

Fig 1
Selected MRI Characteristics and Clinical Symptoms

	Idio	Chiari	Cntrl
# Of Patients	17	17	32
MRI			
Herniation	0	7.6	-
Posterior Fossa Height	24.5	25.5	32.0
Ventral CSF Space	5.7	6.1	12
Dorsal CSF Space	11.5	0.8	19.0

at the base of the skull, through which the spinal cord passes and joins with the brain

idiopathic - due to an unknown cause

posterior fossa - arge opening at the base of the skull, through which the spinal cord passes and joins with the brain

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

syrinx - fluid filled cyst in the spinal cord

ventral - referring to the front of something; can also refer to the bottom

Syrinx Diameter	2.7	5.5	-
Symptoms			
Sensory Loss	94	88	-
Muscle Atrophy	59	65	-
Headache	24	24	-
Ataxia	47	29	-
Scoliosis	53	59	-

Source

Bogdanov EI, Heiss JD, Mendelevich EG, Mikhaylov IM, Haass A. Clinical and neuroimaging features of "idiopathic" syringomyelia. Neurology. 2004 Mar 9;62(5):791-4.