

Key Points

1. Given the numerous symptoms of CM/SM, there is little research on which symptoms improve the most with surgery
2. In reporting the results of 60 patients with CM/SM, researchers analyzed 16 individual symptoms and 15 neurological signs
3. Symptoms and signs were categorized as being caused by Chiari, syringomyelia, or both; in addition, a point system was created to quantify patient improvement
4. All symptoms and signs improved significantly, except for sexual dysfunction (among men), and abnormal reflexes in the arms/hands
5. There was no statistical difference in improvement between CM and SM symptoms; however SM neurological signs improved more than CM signs
6. Half the subjects improved 40%-60%; 20 patients improved less than 40%

Definitions

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 (CM) - 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

dura - thick outer covering of the brain and spinal cord; beneath the dura are the arachnoid and the pia

duraplasty - surgical technique where a patch is sewn into the dura

dysesthesia - an unpleasant, or painful, response to a normal

Brazilian Study Details Which Symptoms Improve With Surgery

One of the limitations of the Chiari and syringomyelia medical literature is that doctors tend to report their treatment results in broad terms; surgery was successful 80% of the time, for example. For a patient suffering from a myriad of symptoms (often 10 or more), such a vague definition of success is of limited value.

In an effort to define surgical outcomes in more detail, Dr. Jose Arnaldo Motta de Arruda, from the Hospital Universitario Walter Cantidio in Brazil, and his colleagues have broken down the surgical results of 60 patients with both Chiari and syringomyelia into discrete symptoms and neurological signs (objective results from a neurological exam). They published their results, on-line, in the journal *Arquivos do Neuro-Psiquiatria* in June, 2004.

Their study encompassed 60 adult patients, with both CM and SM, that were treated between 1982-2000. The average age of the group was 35 years and there 32 women versus 28 men. As is all too common, the average duration of symptoms before surgery was a lengthy 6.2 years. The group underwent the usual battery of tests to verify their conditions, including a neurological exam both before, and at least 6 months after, surgery. A similar surgical technique was used on each patient, namely a decompression with duraplasty and laminectomy. The cerebellar tonsils were either partially or totally removed as part of the procedure as well.

In order to analyze their results, the researchers identified 16 patient reported symptoms (see Table 1) and 15 neurological signs (such as abnormal reflexes, touch response, etc.) and tracked whether the individual symptoms and signs improved in each patient. To examine whether Chiari symptoms and signs improved more or less than ones due to syringomyelia, the doctors also grouped the symptoms and signs into those caused by Chiari, those caused by syringomyelia, and those caused by either or both. Finally, the doctors devised a simple point system to quickly quantify each patient's improvement. For each symptom/sign that a patient had before surgery, the patient was given 0 points if that symptom went away completely, 1 point if it improved, 2 points if it stayed the same, and 3 points if it got worse.

As to be expected, the most commonly reported symptoms before surgery (see Table 1) were muscle weakness, pain in arms/legs, neck pain, and dysesthesia. The most common neurological findings included abnormal reflexes, muscle weakness, abnormal eye movements, and balance problems. Interestingly, the researchers found that more than 70% of the men in the study reported some level of sexual dysfunction, a symptom that is not often discussed in the literature.

In looking at the results after surgery, the group found that nearly every symptom and sign improved to the level of statistical significance except for sexual dysfunction - which only improved in 6 men - and abnormal reflexes in the upper body. In addition to these, and despite a statistical improvement, more than half the patients still suffered from dysesthesia, pain/burning in the limbs, and objectively observed muscle weakness after the surgery, most likely the result of permanent nerve damage caused by the syrinx.

When the researchers compared the improvement in Chiari symptoms versus syringomyelia symptoms, they found no real difference, but when they compared the neurological signs, they found that the syringomyelia signs actually improved more than the Chiari ones.

The results of the point system were interesting as well. The patient scores ranged from 5-55 points with half the patients (30) improving between 40%-60%. Ten patients improved more than 60% and 20 patients improved less than 40%.

While the system this group devised to examine surgical improvement is interesting, it is limited by the fact that it treats all symptoms equally. Slight dysesthesia or weakness in one hand may not limit a patient's quality of life; whereas chronic neck pain, or burning pain in the legs, may have a severe impact on someone's life. It would also be interesting when comparing improvements in Chiari symptoms versus syringomyelia, to look at people who only have Chiari versus people with both. So while this study is definitely a step in the right direction, what is still needed are studies which define success from a patient's point of view, and take into account their quality of life before and after surgery and the overall impact the disease has had.

Table 1
CM & SM Symptoms In 60 Patients

Purple = Chiari
Blue = Syringomyelia
Gray = Either or Both

stimulus; for example pain from being touched lightly

dysphagia - trouble swallowing

laminectomy - surgical removal of part (the bony arch) of one or more vertebrae

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

vertebra - segment of the spinal column, noted as region plus number (C = cervical, T = thoracic, L = Lumbar)

Source

de Arruda JA, Costa CM, de Tella OI Jr. Results of the treatment of syringomyelia associated with Chiari malformation: analysis of 60 cases. Arq Neuropsiquiatr. 2004 Jun;62(2A):237-44. Epub 2004 Jun 23.

Symptom	# With Before Surgery	# With After Surgery
Headache	37	7
Double Vision	21	5
Dysphagia	27	4
Dizziness	39	7
Fainting	34	10
Ear Problems	15	6
Limb Pain	47	15
Muscle Weakness	58	17
Abnormal Sweating	23	13
Pain/burn. in limbs	52	44
Nasal Speech	20	3
Facial Pain	13	3
Sexual Dysfunction (Men Only)	20	14
Problems Walking	40	17
Neck Pain	56	18
Dysesthesia	59	33