Key Points

1. Group reviewed a series of Chiari patients to look for predictors of surgical outcome
2. Examined duration of symptoms, presence of muscle wasting, presence of a dural band, respiratory distress, syrinx, basillar invagination and extent of tonsillar herniation
3. Patients were evaluated - before surgery, at discharge, and 6 months after surgery - using a scale designed to assess CM/SM
4. 6 months after surgery, 62% had good outcomes; 38% had poor outcomes (note, 2 patients died post-operatively)
5. Duration of symptoms, basilar invagination, and respiratory distress were statistically related to clinical outcomes
6. Together these factors were highly predictive of surgical outcome

Definitions

ambulatory - able to walk

appendicular - related to the appendages or limbs

atlanto-occipital membrane (dural band) - fibrous membrane connecting the top vertebra with the edge of the foramen magnum

basilar invagination (BI) - condition, sometimes associated with Chiari, where the C2 vertebra is displaced upward, potentially compressing the brainstem

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 - 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

Researchers In India Look For Predictors Of Surgical Outcome

One of the most pressing questions that every Chiari patient has before undergoing surgery is, "Will the surgery work?" Unfortunately, this question is not always easy to answer, as the research on long-term outcomes is somewhat lacking. With a surgical failure rate as high as 20%, this remains an important question, and doctors and scientists have been working to identify predictors of surgical success for some time (see Related C&S News Stories) with mixed results.

Naturally, one of the major focus areas in the search for predictors is whether a person has a syrinx; however, this is not always a reliable predictor of outcome on an individual basis. Research is beginning to show that the duration of symptoms before surgery is related to outcome (the longer the symptoms existed prior to surgery, the poorer the outcome), but again not every study has shown this to be true. Some researchers have focused in on specific symptoms, while others have looked at the actual shape of the syrinx in trying to find factors that are clearly related to surgical outcome. Interestingly, the factor which originally defined Chiari - the extent of tonsillar herniation - has been shown time and again to NOT be related to outcomes. So despite these efforts, to date there is still no simple, reliable way for a patient to know ahead of time whether surgery will work for them.

Now a group from the Sanjay Gandhi Postgraduate Institute of Medical Sciences, in India, led by Dr. Arora, has used their experiences in treating Chiari and syringomyelia patients to continue the search for accurate predictors of surgical outcome. Specifically, the group looked at whether duration of symptoms, syringomyelia, basilar invagination, respiratory distress, the presence of a dural band, muscle wasting, and the extent of tonsillar herniation were related to surgical outcome in 58 Chiari patients treated between 1991 - 2001. They published their results in the October issue of the journal Neurology India.

The patient group was comprised of 47 men and 11 women with an average age of 27 years. The patients suffered from the usual symptoms associated with Chiari and SM (see Table 1) including pain, motor problems and sensory disturbances. Most of the group (52) had syringomyelia in addition to the Chiari.

After undergoing x-rays and MRI's, the patients underwent similar decompression surgeries; including craniectomy, a laminectomy appropriate for the amount of herniation, and in most cases a duraplasty where the patch was taken from the patient's own body.

In order to measure the surgical outcomes, the researchers modified a disability scale (Klekamp/Samii). Each person was scored on 8 different categories: sensory, muscle strength, gait, urinary/bowel control, appendicular, neck pain, respiratory dysfunction, and lower cranial nerve compression; with a total score ranging from 8 to 32 (8 being the worst and 32 the best possible scores). Each patient was evaluated before surgery, at the time of discharge from the hospital, and 6 months after surgery (it should be noted that only 48 patients were available for evaluation at the 6 month point and that 2 patients died shortly after surgery).

The doctors then classified each patient's overall outcome as either good or poor, with good being defined as the person being able to walk on their own plus an improvement in their disability score. Poor was defined as a person not being able to walk on their own, or a deterioration in their disability score.

Overall, about 60% of the patients experienced good outcomes, while 40% had poor outcomes. At first glance, this result does not seem as good as the success rates generally reported in the US, but it is important to keep in mind that the majority of the patients did have a syrinx, which tends to lower the success rate.

Using statistical techniques, the research group then tried to identify which of the factors were related to - and could predict - the good or poor outcome. They found that duration of symptoms (grouped as <6 months, between 6 months and 3 years, and > 3 years), respiratory distress, and basilar invagination were in fact strongly related to the surgical outcomes. Namely, a longer duration of symptoms, the presence of respiratory problems, and/or basilar invagination were associated with poorer outcomes.

Taken together, the three factors correctly predicted the good outcomes 90% of the time and correctly predicted the poor outcomes 72% of the time. Overall, the factors predicted the outcome 83% of the time. Unfortunately, because of the large discrepancy in the number of people with and without syringomyelia, the group was not able to evaluate whether having a syrinx accurately predicted outcome.

While the outcome measures in this study are somewhat crude compared to what a patient really wants to know (will I be able to work, raise a family, enjoy activities, etc.), it does add to the evidence that duration of symptoms is an important factor in determining outcome after surgery. It also points out the importance of respiratory problems as a symptom and highlights that many Chiari patients have a complex anatomy in that region - basilar invagination being one such example - which can limit the success of decompression surgery.

decompression surgery - general term used for any of several surgical techniques employed to create more space around a Chiari malformation and to relieve compression

dura - tough, outer covering of the brain and spinal cord

duraplasty - surgical technique where the dura is opened and expanded by sewing a patch into it

foramen magnum - large opening at the base of the skull, through which the spinal cord passes and joins with the brain

gait - how a person walks

laminectomy - surgical technique where part of a vertebra is removed

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

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

Source

Arora P, Behari S, Banerji D, Chhabra DK, Jain VK. Factors influencing the outcome in symptomatic Chiari I malformation.
Neurol India. 2004 Oct;52(4):470-4.

Finally, the results also demonstrate the pressing need for earlier diagnoses to improve outcomes and the continued refinement of imaging technologies and surgical techniques to take into account each patient's unique anatomy.

Table 1
Clinical Signs & Symptoms (58 Patients)

Symptom Category	% With Symptom
Pain	34%
Motor	88%
Sensory	79%
Brainstem/Cranial Nerve	48%
Cerebellar	55%
Autonomic	31%
Neck Movement	24%

Table 2
Clinical Outcomes

Outcome	% At Discharge	% At 6 months
Good	60%	63%
Poor	40%	37%

Notes:

Good = able to walk unaided plus an improvement in disability score

Poor = unable to walk unaided, or a deterioration in disability score

There were 58 patients at discharge, but only 48 were available for evaluation at 6 months

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