Key Points

1. Anyone facing Chiari surgery wants to know whether it will work.
2. Most published outcomes studies suffer from limitations but have shown the surgery is effective about 80% of the time.
3. Large study from China involving over 300 surgeries also found that 80% had very good or good outcomes.
4. Two years after surgery, 67% of syrinxes had collapsed or decreased.
5. Like previous studies, outcome was not clearly defined and follow-up times varied.
6. Despite limitations, evidence is accumulating and surgery is successful 80% of the time.

Definitions

aseptic meningitis - inflammation of the covering of the brain, the meninges, which does not involve an infection.
cervical - upper portion of the spinal cord, individual cervical segments are referred to as C1, C2, etc.
craniectomy - surgical procedure where a piece of the cranium, or skull, is removed.
dura - thick, outer covering of the brain and spinal cord.
duraplasty - surgical procedure where the dura is opened and expanded with a patch.
laminectomy - surgical procedure where a portion of one or more bony vertebrae are removed.
retrospective - type of study which looks back in time, often using medical records.
shunt - tube like device used to divert, or drain, CSF.
syrinx - fluid filled cyst in the spinal cord.

Large Study Finds 80% Improve With Surgery

November 30, 2007 -- Anyone facing Chiari surgery wants to know if it will work. Although it is perhaps the most fundamental question regarding Chiari treatment, it has yet to be fully answered. At this point there have been a number of surgical outcome studies published - even some with hundreds of patients - but they all tend to suffer from structural limitations.

For example, most have been retrospective designs, meaning that the research was performed after the fact by using medical records, MRIs, etc. Retrospective designs have a number of weaknesses, one of which is that the data does not always exist to fully answer the question being studied. In other words, since the research was not designed before the patients were evaluated and treated, then the only data available to look at is what was collected on a routine clinical basis. This in turn contributes to somewhat vague and inconsistent outcome definitions.

Post-surgical follow up also continues to be a problem for these studies. Although follow up times are improving, it can take several years to really determine if a Chiari patient improved with surgery and most studies only report follow up times of a year or less. The ones that do go for longer naturally lose touch with patients as time goes by. In this case one has to wonder if it is more likely for people with negative outcomes to change doctors, which would unnaturally bias those that remain towards positive outcomes.

Finally, because the outcome criteria and follow up times in these types of studies are often not well defined, it is difficult to assess the significance and relevance of their findings. Since different studies use different criteria (which are not always well defined) and collect data at different points in time it is also almost impossible to rigorously compare and contrast the research. Replication of results is an underlying principle of the scientific method, but until the bar is raised on the design of these studies, replication will not be possible.

Despite these limitations, and varying definitions of success, in general outcomes studies have shown the same thing. Namely that surgery improves at least the main symptoms of Chiari about 80% of the time. Now a large study out of China (Zhang) reports similar findings for over 300 Chiari patients.

Specifically, the Chinese study involved 316 Chiari patients treated surgically between 1990 and 2006. All of the patients had documented herniations of over 5mm and 236 had syrinxes as well. The surgical technique used varied as determined by the clinical situation of each patient. All patients underwent a craniectomy and a laminectomy of the top two vertebrae. Some patients received more extensive laminotomies depending on the size of the herniation and whether access to a syrinx was required. The dura was opened in a subset of patients and in some patients the syrinx was shunted and drained directly.

As is commonly done, the researchers looked at the outcomes retrospectively using medical records. They chose a simple approach and classified the surgical results as Very Good (complete remission of symptoms or marked improvement), Good (slight improvement of symptoms), Fair (stabilization of symptoms), and Poor (worsening of symptoms). Using this criteria at the time of discharge, 52% of the patients had Very Good outcomes and 29% had Good outcomes (Table 1). Many surgeons would say that either of these outcomes was a successful surgery, so the overall success rate right after surgery was 81%. However, that also means that for 19% of the patients, symptoms did not improve at all immediately after surgery. It is also important to point out that the researchers defined even a slight improvement in symptoms as a Good outcome; it would be interesting to see if the patients in this category agreed that their outcome was good.

In terms of longer term follow up, data was available for a total of 218 patients ranging in time from 5 months to 9 years. The researchers report that among this group the neurological status was improved in 94% of the patients. However, it is not clear what criteria was used to determine this and as mentioned previously there is likely a bias for patients with negative outcomes to switch doctors and thus be lost in terms of follow up.

The authors also reported on follow up MRIs for a subset of the patients with syringomyelia. Images were available for 218 total patients at least two years after surgery. In this group, 35% of the syrinxes had collapsed completely, 32% had decreased in size, 26% were unchanged, and 7% had gotten bigger.

While it is in some ways useful to employ simple definitions of success for outcomes it can also be misleading to patients. To date no one has really studied how Chiari impacts patients' lives after surgery. Did they return to work? Is there marriage stable? Are they able to interact with their kids? Are they able to socialize and engage in recreational activities? Symptoms can improve but still have a significant impact on a person's daily life.

Although this study and the other like it that have come before are interesting, it is time for the Chiari research community to move past retrospective designs with poorly defined criteria.
vertebra - an individual bony segment of the spine

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

cerebellum - part of the brain located at the bottom of the skull, near the opening to the spinal area; important for muscle control, movement, and balance

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

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

Table 1
Surgical Outcomes At Discharge (316 Total Patients)

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Very Good</td>
<td>52%</td>
</tr>
<tr>
<td>Good</td>
<td>29%</td>
</tr>
<tr>
<td>Fair</td>
<td>15%</td>
</tr>
<tr>
<td>Poor</td>
<td>4%</td>
</tr>
</tbody>
</table>

Note: Very good defined as complete remission or marked improvement; Good defined as slight improvement; Fair defined as stable; and Poor defined as worsening symptoms.

Table 2
Long-Term Syrinx Size Outcome (218 Total Patients)

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Collapsed</td>
<td>35%</td>
</tr>
<tr>
<td>Decreased</td>
<td>32%</td>
</tr>
<tr>
<td>Unchanged</td>
<td>26%</td>
</tr>
<tr>
<td>Increased</td>
<td>7%</td>
</tr>
</tbody>
</table>

Note: Based on follow-up MRIs at least 2 years post-op.

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