

Key Points

1. Researchers conducted an international survey of neurosurgeons (76 responded)
2. 25 multiple choice questions using hypothetical cases to examine when and how doctors operate for Chiari
3. Almost all surgeons don't operate if there are no symptoms and no syrinx
4. If there are no symptoms but a syrinx, some surgeons will operate
5. Almost all surgeons will operate if there is a syrinx and progressive scoliosis; about half will operate even if there is no syrinx
6. 76% always open the dura as part of the surgery
7. The most common graft materials reported were pericranial and synthetic

Definitions

asymptomatic - having no symptoms

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

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

decompression surgery - common term for any of several variations of a surgical procedure to alleviate a Chiari malformation

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

dural scoring - surgical technique where a series of cuts are made into the dura, but the dura is not completely opened

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

Survey Shows How Doctors Worldwide Treat Chiari

Surgeons, especially neurosurgeons, tend to have strong opinions; so, in the absence of concrete medical evidence on when and how to operate for Chiari malformations, the neurosurgical community has developed a confusing (for patients) array of surgical techniques and guidelines. As many patients have found first-hand, more often than not, asking two neurosurgeons the same question will yield two different results.

With Chiari, perhaps even more so than some diseases, there seems to be little consensus on topics as fundamental as what defines a Chiari, when to operate, how to operate, when syrinxes form, and a host of other issues. In an attempt to see if there is any consensus, Dr. Edgardo Schijman and Dr. Paul Steinbok, both neurosurgeons, surveyed the international neurosurgical community about when they operate for Chiari and the surgical variations they favor. They published the survey results in February, 2004 in the on-line version of the journal Child's Nervous System.

The researchers designed a 25 multiple choice question survey which also included hypothetical case studies. They distributed the survey to 246 neurosurgeons world-wide and received 76 responses. The responses represented a variety of countries including - but not limited to - the United States, Great Britain, Japan, Argentina, Mexico, the Netherlands, Australia, France, India, and Turkey.

The survey involved four hypothetical case studies, which were used to solicit opinions on when surgery is recommended, plus additional questions on the details of each surgeon's preferred surgical technique. The first two cases (see Figure 1) presented a range from an asymptomatic patient with a significant malformation, but no syrinx, to a patient with headaches, Chiari, and a syrinx. As Figure 1 demonstrates, there is general agreement at the extremes (Case 1 and 2B) - namely don't operate if there are no symptoms and no syrinx, operate if there are symptoms and a syrinx - but not much agreement in the middle grey areas (Cases 1A, 1B, 2, 2A). In fact for Case 2 (headaches, Chiari, no syrinx), the surgeons were almost divided down the middle with 46% saying they would operate, while the rest would either monitor or order further tests.

Interestingly, for the case with an asymptomatic patient with a syrinx (Case 1B), 75% of the surgeons surveyed would operate. The authors point out that this is in contrast with an earlier North American based survey which showed that many surgeons would choose to monitor a situation like this. Additionally, a study of 11 asymptomatic patients with syrinxes which were not operated on showed that only 1 of the 11 eventually required surgery.

The third hypothetical case (3, 3A, 3B) was designed to see when surgery would be recommended in the case of progressive scoliosis. More than half the surgeons would try to stop the progression using decompression surgery even if no syrinx were present, and if a syrinx were present, 97% of the respondents would operate.

The fourth case described an asymptomatic, 12 year old child with Chiari and a long but narrow syrinx. The family does not want surgery, and the surgeons were asked if they would recommend any activity restrictions. Surprisingly, almost half said they would not restrict activities at all, and only 19% said they would recommend avoiding contact sports. More than half the surgeons did say they would explain to the family the risks of not operating.

As expected, the questions dealing with surgical technique generated a wide range of responses. While 95% of the surgeons remove some of the skull as part of the decompression, the amount removed varied widely. When it comes to opening the dura - always a topic of debate - 76% of the respondents said they always open the dura, 20% said they sometimes open the dura, and 1% said they never open the dura. Detailed numbers for another hot topic of debate - whether to shrink or resect the cerebellar tonsils - were not provided. Finally, while many surgeons have begun using tissue from the patient's own body for the dural graft, according to this survey, there is by no means consensus on what material to use. Thirty percent of the surgeons did report they prefer a graft from the patient (pericranial), while 28% prefer to use a synthetic graft, 16% use a graft from a cadaver, and 6% still use material from a cow.

Given the wide range of opinions on so many questions regarding Chiari treatment, it would seem the logical thing for patients to do is to seek out several opinions, understand the reasons behind what each doctor recommends, and find a doctor they agree with and trust.

Figure 1
Response To Operate/Don't Operate Hypothetical Cases

Case #	Would Operate (%)
1	8
1A	28

pericranial - from the periosteum, dense connective tissue which covers the skull

posterior fossa- depression on the inside of the back of the skull, near the base, where the cerebellum is normally situated

scoliosis - abnormal curvature of the spine

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

tonsillar manipulation - surgically removing part, or all, of the cerebellar tonsils

Source

Schijman E, Steinbok P. [International survey on the management of Chiari I malformation and syringomyelia.](#) Childs Nerv Syst. 2004 Feb 14 [Epub ahead of print]

1B	75
2	46
2A	64
2B	90
3	58
3A	85
3B	97

Case Descriptions:

- **Case 1** - 7 year old with no symptoms, 12mm tonsils, no syrx
- **Case 1A**- Same as 1, but with a 2mm wide syrx
- **Case 1B** - Same as 1, but with a 8mm wide syrx
- **Case 2** - 9 year old with headaches, 10mm tonsils, no syrx
- **Case 2A**- Same as 2, but with 2mm syrx
- **Case 2B** - Same as 2, but with 8mm wide syrx
- **Case 3** - 11 year old with progressive scoliosis, 12mm tonsils, no syrx
- **Case 3A**- Same as 3, but with small syrx
- **Case 3B** - Same as 3, but with 6mm wide syrx

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