Key Points

1. Chiari has been linked to many types of eye problems
2. Strabismus occurs when the eyes are misaligned, for example cross-eyed
3. 25 cases of Chiari related strabismus have been reported in the literature
4. It is not known if Chiari causes strabismus or if it is an association
5. Previous research has been mixed on how best to treat
6. Current study describes 12 cases of Chiari related strabismus
7. 9 were successfully treated with strabismus treatments
8. 3 were unsuccessfully treated with decompression surgery
9. Authors caution against neurosurgery if strabismus is the only symptom of Chiari

Definitions

diplopia - double vision

esotropia - type of eye misalignment where the eye(s) turn inward; cross eyed

nystagmus - rapid involuntary movements of the eyes

prism glasses - special type of glasses which utilize a prism to help the eyes work together

scoliosis - abnormal curvature of the spine

strabismus - condition where the eyes are misaligned and do not point in the same direction

strabismus surgery - eye surgery which realigns the muscles that control eye movement in an attempt to align the eyes

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,

Treating Chiari Related Eye Problems

May 20, 2006 -- Eye problems are common with Chiari. The most common, nystagmus, involves rapid, uncontrolled eye movements, and is usually a sign of a neurological problem. In contrast, strabismus, which is a misalignment of the eyes (like being cross-eyed) is most often a problem with the eye muscles, and less often associated with a neurological problem.

In general, when a person looks at an object, both eyes will point at that object. With strabismus this is not the case, and one (or both) eyes will not be lined up properly. Since most of the time strabismus is thought to be a problem with the eye muscle, treatments are directed at correcting this.

Conservative treatments involve patching one eye, or wearing special glasses with a prism built into them which allows the eyes to function as if they are aligned correctly.

Surgery is also a treatment option. Strabismus surgery involves modifying the muscles of the affected eye such that it can move properly and align with the other eye.

While the standard strabismus treatments are well established, doctors are confronted with a dilemma as to how best to treat Chiari related strabismus. It is not know if Chiari can cause strabismus, or if there is just an association between the two. If Chiari does cause strabismus, then logically, treating Chiari - with decompression surgery - may help alleviate strabismus, just like it does other symptoms. However, if the two are just associated, then decompression surgery may not do anything to help strabismus and standard strabismus treatments may be the best approach.

This situation is very similar to scoliosis related to Chiari/syringomyelia. Independent of Chiari, scoliosis treatments include bracing and surgery. However, once doctors identified a link between Chiari/SM and scoliosis, even though the connection was not fully understood, they discovered that treating the Chiari first with decompression surgery helped the scoliosis in many patients. Now, it is becoming standard to only use traditional scoliosis treatments if Chiari surgery fails to improve or stop the curve progression.

However, since strabismus related to Chiari is not as common as scoliosis related to Chiari, the available data has yet to show conclusively what the best treatment approach is. In a study published recently in Binocular Vision & Strabismus Quarterly, Australian eye doctor Lionel Kowal and colleagues reviewed the existing literature on Chiari related strabismus and presented their own experience with 12 patients.

Kowal found 25 cases of acquired strabismus (usually esotropia which means the eye points inward) with Chiari in the medical literature. Many of these patients were under 20 years old, and exhibited other problems, such as headaches, double vision, and abnormal neurological findings.

Treatment and outcome information were available for 21 of the patients (see Table 1). Of those 21, 9 initially received decompression surgery as their treatment, and 12 received standard strabismus treatment (prism glasses or eye surgery). The strabismus resolved for two thirds (6 out of 9) of the patients who initially underwent decompression surgery, and half of the patients who initially received strabismus treatment. Despite the small numbers, these findings have led some researchers to suggest that decompression surgery should be tried first, and strabismus treatment should follow if the condition does not resolve within a year of the initial surgery.

When Kowal's team reviewed their own experiences however, they found much different results. The researchers had treated 12 patients with strabismus who were also found to have Chiari upon MRI. As opposed to the cases reported previously in the literature, very few patients in this group had additional neurological symptoms (other than headaches) and only two had any abnormal findings upon neurological exam.

After consulting with a neurosurgeon, it was decided that 3 of the patients would undergo decompression surgery and the remaining 9 would receive standard strabismus treatment. Interestingly, the strabismus resolved in all 9 of the patients who received strabismus treatment, but did not resolve for any of the three who underwent decompression surgery (see Table 2). In fact, all three patients required additional treatment for their strabismus.

Given the small number of patients in the combined reports, it is difficult to draw strong conclusions at this point about how best to treat Chiari related strabismus. However, the authors of this study do recommend an MRI if strabismus is found with other neurological symptoms or signs, or if the case is unusual. They further state that if strabismus is the only symptom of Chiari, the strabismus may be corrected with standard strabismus treatments, rather than decompression surgery.

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

Source

Source: Kowal L, Yahalom C, Shuey NH.
Chiari 1 malformation presenting as strabismus.
Binocul Vis Strabismus Q. 2006;21(1):18-26.

Given the frequent eye problems associated with Chiari, it is encouraging to see eye doctors become involved in furthering our understanding. To truly understand, and defeat, Chiari will require input from many different disciplines.

Table 1
Previous Research Neurosurgery vs Strabismus Treatment Outcomes

	Effective	Ineffective
Nsg.	6	3
Strabis.	6	6

Notes: Outcomes refer to whether the strabismus was corrected sufficiently; 2 of the 3 patients for whom Chiari surgery was ineffective later underwent strabismus surgery; 4 of 6 patients for whom strabismus treatment was ineffective underwent later decompression surgery

Table 2
Current Study Outcomes, Neurosurgery vs Strabismus Treatment

	Effective	Ineffective
Nsg	0	3
Strabis.	9	0

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