Key Points

1. The cognitive effects of Chiari have not been well established, but there is indirect evidence that it may have a negative effect
2. It has long been noted that spina bifida patients have cognitive impairments, usually attributed to associated hydrocephalus
3. Study examined whether Chiari II associated with spina bifida also contributes to cognitive impairment
4. 46 children with spina bifida and hydrocephalus were given neuropsychological exams
5. Those who also had Chiari II scored significantly lower on Verbal IQ, Performance IQ, and Total IQ
6. Looking at a sub-group of children with Verbal scores over 75 showed that those with Chiari scored lower on visual analysis, verbal memory, and verbal fluency
7. Future research may look at the link between actual anatomical measurements and cognitive scores

Definitions

Chiari II - more severe form of Chiari

cognitive - having to do with conscious, intellectual activities, such as thinking, reasoning, remembering

hydrocephalus - serious condition involving excess accumulation of CSF in the brain area, resulting in increased pressure

IQ - widely used measure of intelligence

myelomeningocele - another name for spina bifida

neuropsychological evaluation - NPE, a series of tests, including standard IQ tests, used to assess the cognitive and emotional impact

Study Identifies Cognitive Impact Of Chiari II

June 20, 2006 -- Since Chiari involves the brain, it is natural to assume that the compression of brain tissue would cause some cognitive problems. While there is abundant anecdotal evidence that this is the case - with many Chiarians referring to a brain fog - there has been virtually no research in this area, so it remains an open question.

Skeptics might point out that the part of the brain affected by Chiari, namely the cerebellum, is thought to control motor functioning, not higher order thought processes. While this was true at one time, more recent research has shown that the cerebellum is likely involved in a wide variety of cognitive functions and activities.

In fact, there is significant indirect evidence that Chiari may in fact be responsible for cognitive impairments. Research involving damage to the cerebellum - not related to Chiari - has shown significant deficits in intelligence tests due to problems such as tumors.

Beyond the cerebellum, Chiari has the potential to affect more than just one part of the brain. The effects of blocking the natural flow of spinal fluid on other parts of the brain are not known. What has been shown, however, is that a long-term increase in intracranial pressure, which is common with Chiari, can have far-reaching cognitive effects.

Interestingly, the cognitive abilities of one sub-group of Chiari patients has been studied extensively, but not because of Chiari. Children born with spina bifida - a birth defect where the spinal cord is exposed - often suffer from cognitive problems (although it should be pointed out that with advances in detection and treatment they are not as severe today). Spina bifida is often accompanied by hydrocephalus, an accumulation of CSF in the brain, and researchers have tended to attribute the cognitive problems associated with spina bifida to the accompanying hydrocephalus.

However, about 30% of children born with spina bifida also have a form of symptomatic Chiari, known as Chiari II. Considered more severe than Chiari I, Chiari II involves parts of the brain beyond the cerebellar tonsils. Given the recent findings that the cerebellum likely plays a large role in higher order thinking, and the high prevalence of Chiari among spina bifida patients, a research team from the Netherlands recently decided to study whether all the cognitive problems associated with spina bifida are really due to hydrocephalus, or whether Chiari plays a role as well.

To determine this, the research team (Vinck et al.) looked at children born between 1988 - 1997 at their hospital with spina bifida and clear MRI evidence of whether they had a Chiari malformation. Out of 78 potential children, 46 ended up participating in the study (note, all participants had had corrective surgery for spina bifida within 5 years of birth).

The researchers divided the children into two groups, 27 who had a Chiari malformation and 19 who didn't (see Table 1). Each child underwent a series of neuropsychological tests to evaluate their cognitive skills. What they found will be published soon in an upcoming issue of the Journal of Neurology, Neurosurgery, and Psychiatry.

When the team analyzed the results, they found that the group of children with Chiari II scored significantly lower on tests of Verbal IQ, Performance IQ, and Total IQ (see Table 2). For example, the average Total IQ score for the group without Chiari II was 94.5 - close to the general population average score of 100 - however, the average score of the group with Chiari II was much lower at 72.6.

While this would seem to be dramatic evidence that Chiari can have a negative effect on cognitive abilities, in this case it could be argued that the children with Chiari II have more severe hydrocephalus, which in turn causes more severe cognitive problems.

To account for this, namely the global cognitive effects of hydrocephalus, the researchers looked more closely at the cognitive profiles of the children, both with and without Chiari II, who had Verbal IQ scores above 75. Presumably, this would help isolate the effects of Chiari, by comparing higher functioning children, whose hydrocephalus - and its effects - were likely mild.

Among the children with Verbal IQ scores above 75, the researchers found significant differences between the Chiari II and no Chiari groups in the specific areas of visual analysis and synthesis, verbal fluency, and verbal memory (verbal fluency is a measure of how easily someone can use words, a sample test would be to name as many fruits as you can in 30 seconds).

While the authors believe that the reduction in visual analysis and synthesis may be due to either hydrocephalus

of neurological diseases and disorders

spina bifida - birth defect where the spinal cord is exposed; often accompanies by hydrocephalus and Chiari II

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

Source

Vinck A, Maassen B, Mullaart R, Rotteveel J. Arnold-Chiari-II malformation and cognitive functioning in spina bifida.
J Neurol Neurosurg Psychiatry. 2006 May 11; [Epub ahead of print]

or Chiari, they point out that the deficits in verbal fluency and memory match the results from studies of other types of damage to the cerebellum, and thus are likely due mostly to the presence of Chiari.

Given the confusing presence of hydrocephalus, it is difficult to say that this study conclusively demonstrates that Chiari can cause cognitive problems; however it certainly adds to the growing amount of evidence that it probably can, and does, have a negative effect on higher-order thought processes.

The scientists in this study suggest that additional research should focus on linking specific anatomical features, as measured by MRI, with specific cognitive deficits. This entire area of research is vital to the Chiari community, which is why it is called out explicitly in both the Conquer Chiari research agenda and the recent call for research proposals, so it is exciting to know that it is receiving at least some attention from the research community.

Table 1
Number and Average Age of Subject Groups

Group	# of subj.	Avg. Age
Chiari	27	10
No CM	19	10
VIQ >75 w/CM	17	9
VIQ >75 no CM	17	9

Note: *Subjects were grouped by Chiari/No Chiari and further divided by those with Verbal IQ's above 75 both with and without Chiari*

Table 2
Cognitive Scores Comparing Chiari to No Chiari Groups

	No CM	CM
Verbal IQ	95.8	80.3
Performance IQ	94.2	68.2
Total IQ	94.5	72.6

Note: *IQ tests are designed so that the median score for a given age is 100, meaning that 50% of people score above 100 and 50% score below*

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