Key Points

1. Patients often wonder if Chiari has cognitive effects
2. Many adults report "brain fog"; some pediatric patients seem to have learning and developmental delays
3. Many doctors dismiss a link between Chiari and cognitive problems
4. However, indirect evidence indicates Chiari may cause cognitive problems
5. This study reports three cases of children with Chiari and abnormal EEG's
6. All three children were found to have blocked CSF flow during surgery
7. EEG's after surgery were normal
8. Authors believe the abnormal EEG's indicate subtle distress of the brain tissue due to blocked CSF flow

Definitions

cognitive - having to do with conscious mental or intellectual activity

EEG - electroencephalograph; device which measures and records the electrical activity of the brain

ICP - intracranial pressure; the pressure of the CSF inside the skull

IQ - a standardized measure of intelligence

IRDA - intermittent rhythmic delta activity, abnormal EEG results which is considered a non-specific indication of a problem

doppler ultrasound - an imaging technology which uses high frequency sound waves; sometimes used during decompression surgery to evaluate CSF flow

cerebellar tonsils - portion of the cerebellum located at the bottom,

Abnormal EEG Results Indicate Chiari's Subtle Effects

April 20, 2006 -- Many Chiari patients ask whether Chiari has cognitive effects. In other words, can it effect critical thinking, problem solving, finding the right words, organizing thoughts, and other types of cognitive functions. This is not an easy question to answer.

On the one hand, many doctors dismiss a link between Chiari and problems with higher order thinking. They focus on the fact that Chiari tends to impact the cerebellum, which traditionally has been thought to control movement and not be involved in higher order thinking.

On the other hand, many adults with Chiari do report cognitive difficulties. These can range from difficulty in articulating thoughts to a general brain fog. In addition, many parents have reported that their children with Chiari may have learning difficulties or developmental delays. Unfortunately, anecdotal evidence such as this can often be misleading, and is not sufficient to say whether Chiari is the cause of the reported cognitive symptoms.

The best way to determine if Chiari is the culprit in cognition would be to evaluate a large number of Chiari patients, both adults and children, with a battery of cognitive and emotional tests to see if on average they score lower than the general population. and if specific problems can be identified. Such tests, known as neuropsycological evaluations (NPE's) are expensive however, and to date research in this area has been essentially non-existent.

Until such research occurs, with no direct evidence of Chiari's role in cognitive problems, indirect evidence is all there is to go on. As this publication has reported on previously, some researchers are beginning to believe that the cerebellum, once relegated to the basement of brain functions, actually plays a large role in a myriad of brain activities.

Studies of children with tumors in the cerebellum region, through NPE's, have demonstrated widespread cognitive deficits. In fact, one researcher has gone so far as to propose the existence of a Cognitive Affective Disorder, where diseases or pathologies in the cerebellum result in a variety of problems with higher-order thinking and emotion.

Another factor to consider when evaluating the possible role of Chiari in cognitive problems is that the effects of Chiari are not always limited to the cerebellum. For most Chiari patients, the malformation blocks the natural flow of cerebrospinal fluid (CSF) between the brain and spinal areas. This blockage can lead to an increase in intracranial pressure (the pressure of the CSF inside the head). Some research has shown that sustained, elevated ICP can lead to long-lasting cognitive problems.

Finally, one has to wonder about the lack of CSF flow itself. CSF bathes the brain and spinal cord and is continuously replaced. What effects does interfering with this natural process have? A recent study out of the University of Siena, Italy (Buoni et al.) may shed some light on this exact question.

In a report posted on-line in March, 2006 in the journal Clinical Neurophysiology, the Italian research team discuss three pediatric Chiari patients treated at their hospital (see Table 1). What makes these patients different is that they were not at first suspected of having Chiari - they didn't have any of the classic symptoms - but rather were being seen for more general problems, such as developmental delays, seizures, and neuromotor delays.

As part of their diagnostic work-up, and before they were found to have Chiari, all three children were given EEG's. An EEG is a device which measures and records the brain's electrical activity through sensors placed on a patient's scalp. In all three cases, the EEG's were abnormal.

Specifically, the tests showed what is called intermittent rhythmic delta activity (IRDA), which is considered a non-specific abnormal result (see Figure 1). In addition, the second patient's EEG showed abnormal spiking as well. The EEG's were recorded at several times for each child and under varying conditions, such as awake, asleep, etc.

Figure1: EEG Of Patient 1 Before And After Chiari Decompression Surgery

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

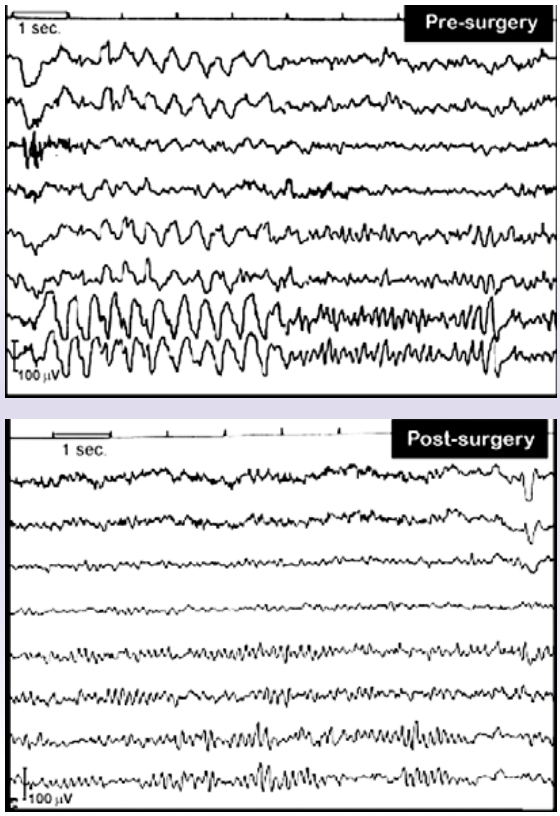
MRI - magnetic resonance imaging; large device which uses strong magnetic fields to produce images of soft tissue inside the human body

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

syrinx - fluid filled cyst in the spinal cord

Source

Source: Buoni S, Zannolli R, Bartolo RM, Donati PA, Mussa F, Giordano F, Genitori L. Surgery removes EEG abnormalities in patients with Chiari type I malformation and poor CSF flow. Clin Neurophysiol. 2006 Mar 15; [Epub ahead of print]



All three children were subsequently given MRI's and found to have Chiari malformations of varying sizes; one child had a syrinx as well. Despite the lack of symptoms directly attributable to Chiari, the physicians decided to operate, and the children underwent Chiari decompression surgery. During surgery, it was noted with ultrasound that CSF flow was severely or completely blocked in each case. Ultrasound also was used to ensure the restoration of normal CSF flow during the course of the procedure.

EEG's were given several times in the year following surgery, and for each child, were completely normal (see Figure 1). The researchers believe that the EEG results indicate a subtle level of distress of the brain tissue due to the lack of CSF flow, which resolved following surgery.

It should be noted that the specific type of EEG result seen, namely IRDA, is thought by some people to represent elevated intracranial pressure. However, the researchers in this study do not believe that is the case here, because the children showed no signs of raised ICP. In addition, evidence has to come light which casts doubt on whether IRDA is linked to elevated ICP at all, further bolstering the author's contention that the EEG findings are likely due to lack of CSF flow.

While it is only three patients, the fact that the EEG's normalized after decompression surgery and the restoration of CSF flow, is pretty compelling. The indirect evidence that Chiari can have a wide-ranging impact on brain function is mounting.

Hopefully, someone will soon look directly at the neuropsychological effects of Chiari so that patients may be properly evaluated and treated. Of course when that research does take place, at least based on this study, it should include EEG's before and after surgery.

Table 1
Description of 3 Patients With Abnormal EEG Results

Age	CM	SM	EEG Before Surgery	EEG After Surgery
12	5mm	N	IRDA	Normal
10	4mm	Y	IRDA w/spikes	Normal
1	8mm	N	IRDA	Normal

Note: IRDA is considered a non-specific abnormal EEG finding

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