Key Points

1. The neuropsychological effects of Chiari have not been studied, however there is a high rate of depression in adults and anecdotally children suffer from cognitive and developmental delays
2. Once thought to only control balance and motor function, the cerebellum is now believed to play a role in higher order cognitive tasks, so it is reasonable that Chiari could interfere with cognitive abilities
3. The neuropsychological effects of posterior fossa tumors have been studied
4. This research evaluated 76 children after being treated for tumors
5. Found that the amount of cerebellar damage was strongly linked to impaired ability as measured by IQ tests
6. Also found that the degree of cerebellar deficit could be measured using a fine motor pegboard test
7. Similar research is required on children with Chiari to identify what, if any, cognitive effects the malformation has and how best to treat deficits

Definitions

ataxia - loss of balance and muscle coordination

brainstem - portion of the brain which connects with the spinal cord; controls many automatic functions such as heart rate, breathing, and swallowing

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

cerebral cortex - part of the brain responsible for conscious

Damage To Cerebellum Affects Cognitive Ability In Children

One of the many aspects of Chiari which has gone virtually untouched by researchers is the psychological effects of the malformation. While it appears that the rate of depression is high among Chiari patients, there is virtually no research studying the effects of having the cerebellum jammed down into the spine on cognitive abilities, or mood and emotion. Anecdotally, one of the most common questions parents of Chiari children ask involves whether Chiari is linked to developmental delays and learning disorders, however it has not been established whether there is a higher rate of learning problems among children with Chiari than their peers.

On the surface, the idea that a Chiari malformation, along with the compression of brain tissue, disrupted flow of spinal fluid, and elevated pressure, might cause problems makes sense. Once thought to be involved just in balance and motor control, the cerebellum is now believed by many to be involved in higher order functioning. In fact, one recent theory posits that the cerebellum plays a regulatory role in nearly all cognitive functions. Functional MRI's, which show activity in the cerebellum during thinking tasks, are providing evidence to support such theories.

The exact role and function of the cerebellum is not yet fully known however, and some theories are still very controversial. One such theory, out of the UK, holds that the cerebellum plays a key role in dyslexia, and that balance exercises, meant to improve function in the cerebellum, improve reading skills. While this might make sense given the growing role of the cerebellum in the brain hierarchy, the evidence to support this theory is hotly contested in the medical community and highlights the importance of direct research on a given subject.

Unfortunately, given the dearth of directly related Chiari research, as with so many aspects of Chiari, we must rely on research in an adjacent area and attempt to apply its findings. On such area is the study of the effects of posterior fossa tumors on children. The posterior fossa is the region where the cerebellum is located and a tumor in that area is similar in some regards to Chiari. There is direct compression of brain tissue, sometimes hydrocephalus develops, intracranial pressure is often elevated, and sometimes a tumor results in an acquired Chiari malformation as the brain tissue is forced out of the skull.

Obviously, a tumor is also different, in potentially important ways. A tumor can involve larger, and different, sections of the cerebellum, and the surgery to remove a tumor involves more direct manipulation of brain tissue than a Chiari decompression. In fact, it is sometimes necessary during a tumor removal to actually cut the vermis, or the center portion of the cerebellum.

Nevertheless, with these significant differences in mind, it is still interesting to look at the work that has been done on the cognitive effects of posterior fossa tumors in children. One such study out of France, led by Dr. Jacques Grill, used neuropyschological evaluations to evaluate intellectual impairments in such children and identify specific predictors of cognitive deficits.

Neuropsychology is the study of the relationship between the brain and behavior. Neuropsychologists often use tests to evaluate a wide range of cognitive function and emotional states in order to identify problems which can then be linked to a physical cause. A thorough neuropsychological evaluation (NPE) encompasses a series of assessments ranging from standard IQ tests, to personality tests, to fine motor control evaluation, and can take several hours.

In Grill's research, 76 children with posterior fossa tumors were given NPE's at least six months after surgical treatment for their tumors. It should be noted that some children also received chemotherapy and/or radiation prior to the surgery.

On average, the IQ scores for the group were lower than the median score of 100 (see Figure 1), with a Verbal IQ average of 87 and Performance IQ average of 76. More striking, however, was that the researchers were able to show that certain characteristics were strongly correlated with the lower IQ scores, namely cerebellar deficits, hydrocephalus, and a cut vermis during surgery. In fact, using statistical techniques, the researchers were able to show that the majority of the variance, or differences, in IQ scores between the children could be predicted by the amount of cerebellar damage (see Fig 2).

To rate cerebellar damage, the researchers used an independent doctor to assess the children for symptoms related to the cerebellum, such as ataxia, nystagmus, and dysmetria. Interestingly, however, they also found that one of the tests in the NPE workup, the Purdue Pegboard test, which requires fine motor control of the hands, was highly correlated with cerebellar damage. Because of this, the score from the Pegboard test was actually an effective predictor of low IQ scores.

While these results are striking, their applicability beyond tumors into the Chiari realm remains an open question.

experience, emotion, thought, and planning

Chiari malformation - 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

cognitive - related to thinking, awareness, and understaning

dysmetria - inability to control the range of muscle movements

intracranial pressure - the pressure of the spinal fluid in the brain

IQ - a crude, but widely used measure of intelligence, remains controversial

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

nystagmus - involuntary, rapid eye movements

posterior fossa - part of the skull, in the back, where the cerebellum is situated

vermis - section of the cerebellum which connects the two hemispheres

Source

Grill J, Viguier D, Kieffer V, Bulteau C, Sainte-Rose C, Hartmann O, Kalifa C, Dellatolas G. Critical risk factors for intellectual impairment in children with posterior fossa tumors: the role of cerebellar damage.J Neurosurg. 2004 Nov;101(2 Suppl):152-8.

Tumors, by their very nature and treatment, would seem able to cause greater damage than Chiari. However Chiari, especially if untreated, does result in direct compression of brain tissue and often elevated intracranial pressure. In addition, some research has shown that elevated intracranial pressure can be more damaging to long-term functioning than lesions themselves.

Obviously, what is needed is direct research on this subject as it relates to Chiari. To begin to understand the neuropsychological impact of Chiari, it would be useful to assess a group of young Chiari patients both before and at various intervals after surgery. It may be that many pediatric Chiari patients would benefit from targeted therapies to improve any cognitive deficits that do exist.

It would also be interesting to see how Chiari patients tend to do on the Pegboard test which was so important in this study and see if it can be used as a simple means to identify the need for an MRI.

Since so little attention has been paid to this aspect of Chiari, the ideas for useful research are many. Given the importance of this subject to the Chiari community, it is time to move past ideas and into action.

Figure 1
Understanding IQ Scores

Range	Category	% of Population within range
55-70	Very Below Average	2%
70-85	Below Average	14%
85-100	Low Average	34%
100-115	High Average	34%
115-130	Above Average	14%
130-145	Very Above Average	2%

Notes: *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; the standard deviation is 15 points, meaning that categories go in ranges of 15 points, thus 68% of the population scores between 85-115 or average; less than 0.1% of people score below 55 or above 145.*

Figure 2
Selected Factors Significantly Associated With IQ Score Variation

Factor	Avg. Verbal IQ	Avg. Perf. IQ
Hydrocephalus	83.4	72.6
Cut Vermis	82.8	69.4
Mild Cerebellar Deficit	88.7	74.6
Moderate Cerebellar Deficit	81.2	70.5
Severe Cerebellar Deficit	63.0	50.6