Key Points

1. The cerebellum, the part of the brain which is displaced in Chiari, has long been thought to be involved only in motor coordination and balance
2. In the last 20 years or so, some researchers have begun to believe that the cerebellum plays a larger role in cognitive processes
3. Scientists use functional imaging and examine people with damage to study the role of different brain regions
4. In 1998, study was published which proposed the Cerebellar Cognitive Affective Syndrome
5. Looked at 20 patients with damage limited to their cerebellum
6. Testing revealed problems with executive functions, visuospatial tasks, language problems, and personality changes, including flat affect and inappropriate behavior
7. Patients studied likely had more direct damage to the cerebellum than Chiari patients, but it does indicate that the cerebellum is involved in higher-order thinking
8. Recently, even more evidence supporting this has come to light

Definitions

affect - how a person acts related to their mood and emotions

agrammatism - not using connecting words in speech

anomia - trouble finding the appropriate word

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

cerebellitis - inflammation of the cerebellum usually due to a viral infection

cerebellum - part of the brain located at the bottom of the skull,

What Is The Cerebellar Cognitive Affective Syndrome

October 15, 2005 -- The cerebellum, that part of the brain which is of intense interest to the Chiari community, is well known in its role as the conductor of movement and coordination. For 100 years, neuroscientists believed that movement was not only the primary job of the cerebellum, but that it was its only job. Over the years, there have been isolated reports of patients with damage to their cerebellum exhibiting problems with language and higher-order thinking, but these received little attention from the medical community and were generally dismissed.

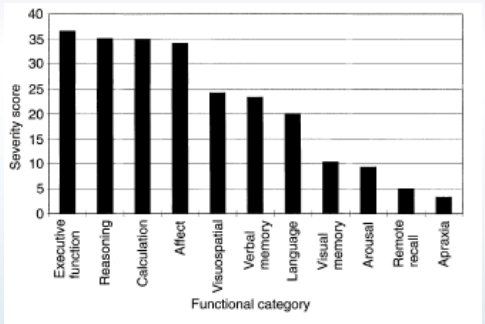
Then, about 20 years ago, as occurs regularly in science, some researchers began to rethink this dogma, and began to explore whether the cerebellum actually controlled more than physical movement. Utilizing advances in imaging technologies which could show what brain regions were active during functional tasks, along with more finely tuned neuropsychological tests, a small body of evidence began to emerge which hinted that the traditional thinking about the cerebellum was wrong, and that it actually played a role, potentially a vital one at that, in many higher level cognitive processes.

Old ideas die hard, especially in science, and while today the evidence continues to mount, the debate on what role the cerebellum plays is far from over; twenty years ago, it was just beginning. One of the most powerful pieces of evidence in favor of a larger role for the cerebellum came from Dr. Jeremy Schmahmann and a colleague at Harvard Medical School. In 1998, in the journal Brain, Schmahmann published a study which tried to address some of the concerns at the time, by thoroughly examining 20 patients with diseases limited to their cerebellums and showing the clinical implications of the cerebellar damage. In describing his results, he coined the term Cerebellar Cognitive Affective Syndrome.

The study, which became fairly widely cited (244 citations according to Google Scholar), was conducted over a seven year period. Patients with a disease involving the cerebellum, such as a stroke or cerebellitis, were given a neurological exam, bedside mental evaluations, MRI's, EEG's, and extensive neuropsychological testing. People with lesions in the brain outside the cerebellum, major neurological diseases, head trauma, history of drug/alcohol abuse, or a number of other medical conditions were excluded. In other words, the researchers went to great lengths to find people who appeared to only have a problem confined to their cerebellum.

In performing the bedside mental state evaluations, the team used well-established clinical methods to rate the level of impairment from mild (1) to severe (3) in several areas. To compare the functional areas, they then calculated a total severity score by adding the individual scores and creating a ratio versus the worst score possible for the entire group. They found that the most impaired functional areas involved executive function, reasoning, calculation, affect, visuospatial tasks, verbal memory, and language (Figure 1).

Figure 1
Bedside Mental State Evaluation Impairment Results By Functional Category



Notes: *The most impaired functional areas were Executive Function, Reasoning, Calculation, Affect, Visuospatial, Verbal Memory, and Language*

Neuropsychological testing was administered some time later, and in some cases at a follow-up visit as well. Not surprisingly, these tests revealed similar results, with the IQ scores, executive functioning, reasoning, visuospatial, attention, and visual reproduction scores all showing very significant deficits as compared to normal. In addition, a couple of other tests revealed less, but still significant, differences as well.

To better highlight the clinical picture, the Harvard team also described a couple of the cases they studied. One woman, a 22-year old college student, had had a tumor removed from her cerebellum. After the surgery, her

near the opening to the spinal area; important for muscle control, movement, and balance

cerebral cortex - part of the brain responsible for conscious 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 understanding

EEG - electroencephalography, test which records the brain's electrical activity

executive function - in cognitive theory, system which controls other cognitive actions; involves high-level functions such as planning, prioritizing, abstract thinking, etc.

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

vermis - section of the cerebellum which connects the two hemispheres

Source

Source: Schmahmann JD, Sherman JC. The cerebellar cognitive affective syndrome. Brain. 1998 Apr;121 (Pt 4):561-79.

personality changed dramatically. She would behave inappropriately, including undressing in the hallways, and she had become very soft-spoken with virtually no affect. She had tremendous difficulty with many of the tests and continued to behave in a child-like manner. She improved slowly over a period of months, and was able to enter a remedial program at a junior college two years after her surgery.

In analyzing and discussing their results as a whole, the authors believe that damage to the cerebellum results in what they call Cerebellar Cognitive Affective Syndrome. This syndrome is characterized by:

1. "Disturbances of executive function. This includes deficient planning, set-shifting, abstract reasoning, working memory, and decreased verbal fluency."
2. "Impaired spatial cognition, including visuospatial disorganization and impaired visuospatial memory."
3. "Personality change, characterized by flattening or blunting of affect, and disinhibited or inappropriate behavior."
4. "Linguistic difficulties, including dysprosodia, agrammatism, and mild anomia."

They conclude that in general these patients experience an overall reduction in their intellectual ability.

So what does this mean for Chiari patients? Fist, care must be taken in interpreting this study as it relates to Chiari. In Chiari 1, the bottom of the cerebellum, the tonsils, are what is known to get crowded. Post-surgical examination of tonsils that were removed has shown that this brain tissue is generally abnormal. However, it is not clear what type of damage might occur to the rest of the cerebellum. The majority of patients in this study had suffered strokes, resulting in extensive damage to large areas of the cerebellum.

In addition, before we jump to the conclusion that Chiari causes cognitive problems, we must keep in mind, as Dr. Bland explains in her column, Let's Think About Thinking - The Cognitive Effects Of Chiari, there can be several reasons why someone with Chiari may experience cognitive difficulties. Chronic pain, medications, depression, and unrelated conditions all must be considered as possible contributors to any cognitive problems.

Given this, what can be taken from this research, and other similar research, is that there is growing evidence that the cerebellum plays a vital role in many cognitive functions and tasks. Therefore, there is a strong theoretical basis for exploring, through rigorous research, what the cognitive effects of Chiari may be.

Figure 2
Selected NPE Tests Which Showed The Most Statistically Significant Impaired Results

1. Intellectual Functioning - Full-scale IQ, Verbal IQ, Performance IQ
2. Executive Functioning - Word Association, Animal Naming
3. Reasoning - Picture Arrangement
4. Visuospatial - Complex Figure Copy, Block Diagram
5. Attention - Digit Symbol
6. Visual Reproduction

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