Healing Gets In The Way; Rapid Chiari Onset

Case Studies is a feature designed to highlight interesting patient cases reported in the research. Given the lack of knowledge about CM/SM, much of the published research comes in the form of case studies - doctors describing one or two patients they have seen and treated - as opposed to rigorous scientific studies. While this type of publication doesn't advance the scientific cause as much, it does give us a window into some of the issues surrounding CM/SM, including lasting side effects and related conditions. And hopefully, some of our readers will say, "Hey, that's just like me!" and know they are not alone in what they are going through.

CASE 1: Rapid Development of Chiari I Malformation In An Infant With Seckel Syndrome and Craniosynostosis

Reported In: Journal of Neurosurgery. May, 2003.
Doctors: Timothy Hopkins, M.D., Stephen Haines, M.D., University of South Carolina
Patient:

- 3-month old male infant with many abnormalities, including growth retardation, Seckel syndrome (see Side Bar) and craniosynostosis
- Baby was taken to the ER after having trouble breathing at home for an extended period of time
- MRI showed problems, but no tonsillar herniation
- A resistant Staph infection was identified and treated and the baby was sent home
- Several days later, the baby returned to the ER again having trouble breathing
- MRI this time revealed a Chiari malformation extending beyond C-2 and compressing the brainstem
- Baby underwent decompression surgery, but unfortunately passed away several weeks later from other causes

Observations:

- Authors identified 22 cases in the literature, documented by images, where an initial image showed no Chiari and a later image showed a malformation
- Time between images ranged from as short as 11 days to as long as 18 years
- Acquired Chiari has been associated with placement of lumboperitoneal shunts to treat hydrocephalus, traumatic lumbar punctures, and craniosynostosis among others causes

Ed Note: *This sad case illustrates that a Chairi malformation is not always congenital. While this was the prevailing thought for many years, some researchers are now thinking about what role trauma plays in either causing a malformation or turning an otherwise benign malformation into a symptomatic one. For those who developed symptoms in adulthood, it is interesting to speculate whether the malformation was always there.*

CASE 2: Reformation Of The Posterior Atlanto-Occipital Membrane Following Decompression Surgery

Reported In: Pediatric Neurosurgery (Case Report). April, 2003
Doctors: Dr. Tubbs, Dr. Wellons, Dr. Oakes, Dr. Blount; University of Alabama at Birmingham
Patient:

- 5 year old male diagnosed with Chiari and a large syrinx but no significant neurological problems, other than a seizure
- Underwent decompression surgery with no complications and appeared to recover
- 3 months later, reported some neurological symptoms
- MRI showed the syrinx was larger and there was crowding around the cerebellum
- Exploratory surgery revealed that the atlanto-occiptal membrane, despite being cut during the first surgery, had regrown and was constricting the craniocervical junction. The membrane was removed

Definitions

brainstem - lowest part of the brain, connects with the spinal cord

cerebrospinal fluid (CSF) - clear liquid in the brain and spinal cord, acts as a shock absorber

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

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

craniocervical junction - area where the skull meets the spine

craniosynostosis - conditions where the sutures of the skull close prematurely, resulting in a misshaped skull

dura - thick outer layer covering the brain and spinal cord

decompression surgery - common term for any of several variations of a surgical procedure to alleviate a Chiari malformation

foramen magnum - opening at the base of the skull, through which the spinal cord passes

posterior atlanto-occipital membrane - fibrous membrane connecting the back of the top vertebra with the edge of the foramen magnum

Seckel Syndrome - rare, genetic condition which involves growth retardation and sometimes mental retardation

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

syrinx - fluid filled cyst in the spinal cord

vertebra - segment of the spinal column

- 3 months later, symptoms had resolved and MRI revealed significant decrease in syrinx size

Observations:

- The posterior atlanto-occipital membrane is made of tough, fibrous, connective tissue
- It is more than 4 times as strong as the dura tissue
- Once thought to add to neck stability, the membrane is now thought to contribute very little to stability
- Surgeons performing a reoperation should evaluate the membrane to see if it is causing constriction

Ed Note: *This case illustrates another reason why reoperations are necessary in some cases. Some researchers speculate that the membrane becomes tougher and more stiff in some adults and contributes to the compression of the area.*