Key Points

1. Syringobulbia involves the formation of a syrinx in the brainstem
2. Reports indicate that between 1-10% of Chiari related syringomyelia patients also have syringobulbia
3. Study used morphometric analysis to see if skull base geometry could explain why some patients develop syringobulbia
4. Reviewed 189 cases and identified two with syringobulbia
5. Despite numerous measurements, could not find a single significant difference between syringobulbia, syringomyelia with out syringobulbia, and Chiari only patients
6. Could be that they missed the measurement that is the key or that the mechanism is something else entirely
7. Syringobulbia patients offer an interesting opportunity to explore and test syrinx theories

Definitions

brainstem - part of the brain which connects to the spinal cord and controls basic functions such as breathing

foramen magnum - opening at the base of the skull through which the brain and spinal cord meet

morphometric - in this article, refers to measuring dimensions of the skull and brain

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

scoliosis - abnormal curvature of the spine

syringobulbia - condition, most often associated with Chiari and syringomyelia, where fluid collects in the brainstem

Syrinx In The Brainstem Raises Questions

March 31st, 2009 -- As if having a syrinx in the spine weren't bad enough, imagine having one in your brainstem (Figure 1). The brainstem is the part of the brain that connects to the spinal cord and controls many basic bodily functions, such as breathing and heart rate. When a syrinx forms in the brainstem it is called syringobulbia. Published reports of patient series indicate that this occurs in anywhere from about 1% of syringomyelia cases to as many as 10%.

In an effort to understand why some people develop syrinxes in their brainstem, a well published group from the University of Alabama, Birmingham (Tubbs et al.) used morphometric analysis to see if the skull shape of patients with syringobulbia was different in any way from patients with spinal syrinxes and Chiari patients with no syrinxes. They published their results on-line in the journal, Child's Nervous System.

Morphology refers to studying the form and structure of something. Many types of science, such as biology, geology, and even astronomy, have morphology branches which quantify and classify the structure of everything from plants, to rock formations, to galaxies. In the Chiari world, morphometrics has been an active area of research for several years, ever since publications started emerging which showed Chiari patients tend to have smaller posterior fossas than average. In these types of studies, researchers use MRIs to take numerous distance and angle measurement of the skull base and then compare different groups. While certain research publications have definitely shown that on average some Chiari patients have small posterior fossas, this is not universal, and not all research has found this to be true. Currently, the utility of these types of measurements is still being evaluated.

In this study, the UAB researchers hypothesized that patients who develop syringobulbia have a different skull base anatomy than patients who have spinal syrinxes (but no brainstem syrinx) and Chiari patients with no syrinxes. To test their hypothesis, the group reviewed the records of 189 children and took numerous morphometric measurements from each of their MRIs.

Out of the entire group of children, they found only two cases of syringobulbia, which represented about 1% of the patients. Both cases had significant herniations (7mm and 10mm) and symptoms such as headaches and scoliosis. Interestingly, despite the syrinxes in their brainstems, neither patient had symptoms or neurological signs, such as breathing issues or cranial nerve involvement, directly attributable to those syrinxes. In each case, surgery resulted in a reduction in the size of both the brainstem syrinxes and the spinal cord syrinxes.

While the clinical outcomes were good, the results of their research failed to support their hypothesis. Specifically, despite taking numerous measurements, the researchers failed to find a single morphometric measurement that was significantly different between the patients with syringobulbia, the patients with syringomyelia but no syringobulbia, and the Chiari patients without either.

Although the authors of this study do not directly discuss this in their publication, there are several possibilities as to why they didn't find anything [Ed. Note: The following is based in part on a discussion with a syringomyelia expert not affiliated with this research]. One possibility is that the research group simply did not find the morphometric measurement, or combination of measurements, that leads to the development of syringobulbia. This is made more likely by the fact that they only had two syringobulbia patients to work with. A second possibility is that the mechanism involved has nothing to do with skull base geometry, but rather is related to something different entirely, such as the variations in the tissue properties between individuals. A third possibility is that in patients such as these, and in patients who appear to have multiple syrinxes that are isolated from each other, it could be that current MRI technology is not powerful enough to show if the syrinxes are actually linked somehow within the tissue, and because of this, they appear to be separate from each other.

Whatever the underlying reason for the development of syringobulbia turns out to be, using these cases to test theories is a very interesting, and useful, approach. As the authors do point out, most syrinx theories focus on the blockage of CSF at the foramen magnum, and how the development of syrinxes in the brainstem fits into those models, is not at all clear.

Figure 1: MRI Of Syringobulbia Patient

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

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

syringomyelia - condition where a fluid filled cyst forms in the spinal cord

Source

Morphometric analysis of the craniocervical juncture in children with Chiari I malformation and concomitant syringobulbia. Tubbs RS, Bailey M, Barrow WC, Loukas M, Shoja MM, Oakes WJ. Childs Nerv Syst. 2009 Feb 13. [Epub ahead of print]

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