Special Report: 01.09

Key Points

1. There were149 English language studies published in 2008 involving Chiari and/or syringomyelia
2. 48% of those were either Case Studies or mentioned Chiari incidentally
3. There were quite a few studies focused on surgical outcomes (14)
4. Restless Leg Syndrome was tied to Chiari
5. The relationship between Tethered Cord Syndrome and Chiari remains controversial.
6. Some experts are pushing for adoption of a new nomenclature for Chiari, based on the underlying cause

Definitions

case study - type of research which focuses on one subject, or person, as opposed to a group of subjects

hypothesis - a proposed idea, not yet tested, about how or why something works

prospective - type of study where the experiment is designed before the data is collected

PubMed - database maintained by the National Library of Medicine, includes over 15 million citations for biomedical articles

randomized - technique used in a scientific study where participants are randomly assigned to one of two groups; used to control the effects of age, gender, etc. on the study outcome

retrospective - type of scientific study which looks back in time, often at medical records

review - type of publication which summarizes the current knowledge on a given subject; usually does not provide new data or hypotheses

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

Year In Research: 2008, The Song Remains The Same

Unfortunately, when it comes to Chiari research, in large part the song remains the same. In 2008, there were 149 English language publications on Chiari and/or syringomyelia (Figure 1). Although this is a slight decrease from the previous year, when studies where CM/SM were not the primary focus and case studies which involve only a couple of patients are removed, the adjusted total (78) (Figure 2) actually represents a slight increase over 2007. This means that even though the total quantity of research dipped, the number of quality studies increased slightly. However, both numbers remain ridiculously low as compared to diseases such as multiple sclerosis.

Table 1
2008 PubMed CM/SM Citations By Subject Type (149 Total)

Subject	# of Citations
Total	149
Incidental Reference	13
Case Study	58
Outcomes	14
Surgical Technique	8
Theories	3
Imaging/Diagnosis	7
Symptoms	11
Related Conditions	2
Chiari II/Spina Bifida	12
Animal	9
Morphological	2
Reviews	2
Scoliosis	2
PTS	3
Acquired	2

Notes:

- Incidental refers to a study which mentions Chiari or syringomyelia in passing, but CM/SM is not the focus of the publication
- Case studies refer to publications which report on 3 or fewer patients and are mostly descriptive in nature

Table 2
Yearly CM/SM Research Citations (2004-2008)

	'08	'07	'06	'05	'04
Total English Language Citations	149	161	131	137	122
Adjusted Total	78	72	52	39	53

Note: Adjusted Total refers to the Total Number of Citations minus the Incidental and Case Study citations

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Interestingly, among the limited pool of Chiari research, outcome studies, symptoms, and imaging studies represented more than 40% of the work. In addition, there were the usual publications on animals, skull dimensions (morphology), and spina bifida.
Some of the highlights from the 2008 research include:

- A continued, although controversial, focus on the potential relationship between Tethered Cord Syndrome and Chiari
- Strong scientific data that a subset of Chiari patients also have connective tissue issues which can affect

so named because of their shape

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

syrinx - fluid filled cyst in the spinal cord

tonsillar herniation - descent of the cerebellar tonsils into the spinal area; often measured in mm

Source

PubMed search with keywords Chiari, syringomyelia and limit of publication date between 1/1/08 and 12/31/08. Duplicate results were eliminated manually. Citations were categorized by the editor. Foreign language publications were excluded.

their treatment needs

- Studies linking migraines, childhood hearing loss, and restless leg syndrome to Chiari
- A detailed look at the risks which Chiari surgery can pose in terms of cervical instability and the treatment required to stabilize it
- In a positive move, some outcome studies have begun to track symptom recurrence over time; which unfortunately showed that symptoms due tend to come back for some people

Not to pat ourselves on the back too hard, but perhaps the most significant event in Chiari research in 2008 was the NIH sponsored Conquer Chiari Research Conference. The event was extremely well attended and generated tremendous discussion. In addition, it brought Chiari into the awareness of NIH, which hopefully will result in more funding.

One of the highlights of the conference was a presentation by Dr. Milhorat, in which he suggested that Chiari patients could be categorized by the the underlying cause of the Chiari: small posterior fossa, tethered cord, connective tissue issues, CSF issues, etc. Conquer Chiari has spoken out in favor of such an approach in the past and wholeheartedly supports this type of progress.

Working groups at the conference generated ideas ranging from a patient registry to a large genetic study to a newsletter for general neurosurgeons about Chiari.

Although the progress often seems slow, and the answers can not come soon enough, progress is being made and there is a growing hope that one day we will Conquer Chiari.

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