

Key Points

1. 2006 was not much different from the previous two years in terms of quantity and quality of published research
2. There were 131 English language publications
3. The majority of these (79) were either incidental references or Case Studies
4. Chiari/SM continues to lag far behind other diseases in quantity of published research
5. Conquer Chiari is working to improve the situation through our Research Agenda, Research Conference, and Research Funding

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

Multiple Sclerosis - serious, chronic, neurological disease, where for reasons unknown, the covering of nerve fibers are destroyed, disrupting the normal nerve activity

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

Year In Research: 2006, Same As It Ever Was

It appears that some thing in the Chiari community never change. At least that's the case with the research which was published in 2006. An analysis by Chiari & Syringomyelia News shows that in both quantity and quality, the Chiari (and syringomyelia) research has essentially remained static for the last 3 years.

The results from a PubMed search revealed 131 English language Chiari and SM research publications in 2006. This represents a slight 4% decrease from 2005 (137 citations), which was a slight increase from 2004 (see Table 1). More troubling however, is that much like the previous two years, the majority of the 131 citations were either incidental mentions of Chiari, or simple Case Studies describing one or two patients.

Table 1
2006 PubMed CM/SM Citations By Subject Type (131 Total)

Subject	# of Citations
Total	131
Incidental Reference	27
Case Study	52
Chiari II	9
Theoretical	6
Symptoms	6
Associated Diseases	6
Surgical/Results	6
Animal	5
Syringomyelia	5
MRI/CSF Flow	4
Scoliosis	2
Genetic	1
Epidemiological	1
Miscellaneous	1

- 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

Specifically, 79, or a full 60% of the 2006 publications fall into this category. That means that only 52 publications focused on CM/SM which were not Case Studies. This is an incredibly low number of research publications for a disease which likely affects more than 300,000 Americans.

A couple of categories were able to buck the trend however. Nine studies were published on Chiari II, a significant jump from 2005. Similarly, six studies of a theoretical nature were published, compared to 2 the previous year. However, the overall numbers are so low, that minor jumps in some categories is to be expected.

Despite the limited amount of research, important progress was made in a number of areas:

1. A number of studies were published regarding the concept of Tethered Cord Syndrome, whether it can exist without being seen on MRI, and what relation it might have to Chiari.
2. Two studies were published which examined the neuropsychological effects of Chiari II and found that indeed children with Chiari II are more likely to have cognitive problems.
3. The relationship between pseudo-tumor cerebri and Chiari was further elucidated by a study which found a high rate of Chiari among PTC patients. This adds to the notion that the hydrodynamics of blood and CSF in the brain and spine are critical to understanding Chiari.
4. Researchers used advanced imaging to study why some people with SM develop central neuropathic pain and other don't.

or hypotheses

cerebellar tonsils - portion of the cerebellum located at the bottom, 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 1/1/06 and 12/31/06. Duplicate results were eliminated manually. Citations were categorized by the editor. Foreign language publications were excluded.

5. By screening families with multiple members affected by Chiari, researchers at Duke University for the first time identified a possible gene that may play a role in Chiari.

While these advances are exciting, the pace of Chiari research, as evidenced by the last three years, is simply not fast enough. As this publication has pointed out before, the US government earmarks tens of millions of dollars for research into diseases such as MS, but virtually nothing for Chiari and syringomyelia.

Table 2
Yearly CM/SM Research Citations (2004-2006)

	'06	'05	'04
Total English Language Citations	131	137	122
Adjusted Total	52	39	53

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

Conquer Chiari is trying to address the research situation in three ways. First, we have begun to award research grants. The best way to spark research is by supplying funds. Second, we are organizing the first Conquer Chiari Research Symposium for June, 2007. This will be a one day professional workshop which will bring together a number of the top physicians and scientists in the field to present their work in a collaborative environment.

Finally, Conquer Chiari will use both our grant program and the research conference to promote our Research Agenda. The Conquer Chiari Research Agenda is intended to maximize the impact we can have and help coordinate the research that is already occurring in this space (see below).

Conquer Chiari Research Agenda: A Roadmap For Success

Goal #1: Reduce the average time to an accurate diagnosis to less than 2 years from time of first symptoms.

Objectives:

- Develop a standard, simple, objective definition and test of symptomatic Chiari
- Enable the introduction of new technologies, such as inexpensive, portable imaging, which will reduce the barriers to diagnosis

Goal #2: Develop an effective, widely adopted, and minimally traumatic standard of care.

Objectives:

- Design, and encourage the adoption of, a standard outcome measure, such that the results from different studies can be compared and combined
- Establish whether the surgical variations that currently exist have a significant effect on long-term patient outcomes, and further develop a standardized surgical approach
- Encourage the development of minimally invasive surgical techniques
- Pursue non-surgical treatment approaches which don't just address symptoms, but are targeted at the core problem(s)

Goal #3: Minimize the impact that Chiari has on the quality of life of patients.

Objectives:

- Develop, and encourage the adoption of, a Chiari Impact Measure, which takes into account patient focused issues such as career, family, economics, recreation, and socialization
- Understand, and develop treatments for, the neuropsychological effects of Chiari, including both cognitive and emotional manifestations
- Develop widely accepted protocols for physical, occupational, and other types of therapies designed to maximize functional capabilities
- Enable the development of innovative technologies and treatments targeted at the neuropathic pain and loss of function associated with Chiari

Goal #4: Understand the pathophysiology, natural history, and epidemiological characteristics of Chiari.

Objectives:

- Establish, with reasonable accuracy, the incidence and prevalence of Chiari and Chiari related syringomyelia
- Characterize, and quantify, the Chiari experience, such as average age of diagnosis, time to diagnosis,

number of doctors seen, major symptoms, etc.

- Develop a sound theoretical model for the pathophysiology of Chiari, which explains how symptoms develop, and will enable predictions about who needs surgery, who will develop syringomyelia, etc.

- Identify and characterize the genetic basis of Chiari

Chiari research has been limping along at a snail's pace for the last 3 years. Conquer Chiari is trying to change that, and with the help of the entire Chiari community, we can make sure that Chiari & Syringomyelia News has a lot to write about in the years ahead.

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