

Key Points

1. 2005 was not much different from 2004 in terms of quantity of research published
2. There were 137 English language publications
3. The majority of these were Case Reports (72)
4. Chiari/SM continues to lag far behind other diseases in quantity of published research
5. May be due to surgical nature of treatment

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 or hypotheses

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

Year In Research: 2005

When it comes to Chiari research, it appears that history does repeat itself...over and over again. Much like the year before, 2005 saw what could only be described as a discouragingly low number of research publications, once again dominated by Case Reports.

According to PubMed, there 137 publications in 2005 which at least mentioned Chiari and/or syringomyelia (see Table 1). Of these, 26 were focused on other topics and only incidentally referred to CM/SM. Setting these aside, a whopping 72 of the remaining 111 publications were Case Reports. This means that 65% of all publications where Chiari and/or SM was the main subject were actually just descriptions of one or two patients. Unfortunately, Case Reports do little to advance the scientific knowledge of the disease, because one or two patients is not enough to draw any conclusions from.

Table 1
2005 PubMed CM/SM Citations By Subject Type (137 Total)

| Subject | # of Citations | Difference From 2004 |
|------------------------|----------------|----------------------|
| Total | 137 | +15 |
| Incidental Reference | 26 | +6 |
| Case Study | 72 | +23 |
| Surgical/Results | 9 | -2 |
| Animal | 5 | -1 |
| Reviews | 5 | 0 |
| Non-Chiari SM | 5 | +1 |
| MRI/CSF Flow | 4 | -2 |
| Chiari II | 3 | -1 |
| Theoretical | 2 | -2 |
| Posterior Fossa Volume | 2 | 0 |
| Symptoms | 1 | -3 |
| Scoliosis | 1 | -2 |
| Associated Diseases | 1 | 0 |
| Miscellaneous | 1 | - |

- Incidental refers to a study which mentions Chiari or syringomyelia in passing, but CM/SM is not the focus of the publication
- Reviews refer to papers which simply summarized current knowledge, but did not report any new data or ideas
- Case studies refer to publications which report on 3 or fewer patients and are mostly descriptive in nature

There continues to be a conspicuous absence of the most rigorous types of scientific studies, randomized controlled trials. There was one prospective, randomized trial which compared the use of synthetic dural grafts versus tissue taken from the patient; however the publication was in Chinese.

For better or worse, the Chiari literature continues to be dominated by physicians reporting on their experience with patients and retrospective studies. Despite this, there were a number of interesting publications during the year:

1. A study of over 100 Chiari patients found that about 80% reported a significant improvement in their quality of life after decompression surgery
2. Although it was not the main focus of the research, an article in the prestigious journal, Nature, used the results from tracking the development of individual fetal cells to hypothesize that Chiari is due to an error in the development of the clivus (a bone in the lower part of the skull), which doesn't form properly. In addition, the article cited a specific gene which may be involved.
3. Advanced MRI techniques were used to show that compliance, which refers to how flexible the covering of

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

Source: PubMed search with keywords Chiari, syringomyelia and limit of publication date between 1/1/05 and 12/31/05. Duplicate results were eliminated manually. Citations were categorized by the editor. Foreign language publications were excluded.

the brain/spine is, is abnormally low in people with Chiari and improves with successful decompression surgery

4. Research continued to show that Chiari patients have abnormally small posterior fossa skull regions, the area in the back of the skull where the cerebellum and brain stem are situated.
5. A speculative, and somewhat controversial, article proposed a new surgery to treat Chiari. Namely, cutting the filum terminale at the base of the spine. While the research was not rigorously designed, if proven to be effective, this approach would represent a major deviation from the current accepted practice

So while there were some advances, in some ways, 2005 was not as productive as the year before. Despite a 12% increase in the total number of publications, there were fewer works on surgical results, animal studies, CSF flow, Chiari II, new theories, symptoms, and scoliosis in 2005 than in 2004. In fact, the slight increase in publications was almost completely due to a dramatic increase in the number of Case Reports.

Not surprisingly, Chiari continues to lag far behind other diseases in both quantity and quality of research. Multiple Sclerosis, which in many ways is similar to Chiari, had nearly 2,000 publications in the medical literature in 2005 (see Table 2). Similarly, diseases which affect fewer people but with more serious consequences, such as ALS and Huntington's, saw 3-5 times as much research activity as Chiari.

Table 2
Number of 2005 Citations For Various Diseases

| Disease | # of Citations |
|--------------------|----------------|
| Multiple Sclerosis | 1,972 |
| Parkinson | >1,000 |
| Hydrocephalus | 648 |
| ALS | 508 |
| Huntington | 369 |
| Spinal Stenosis | 148 |

While it is not entirely clear why Chiari continues to essentially be ignored year after year, several factors may play a role. Clearly, awareness and advocacy are a big part of the equation. Multiple Sclerosis has a long history of successful advocacy groups who have funneled enormous amounts of money towards their cause.

Another issue may be the fact that there are no good estimates of how many people suffer from Chiari and how it impacts them. Without this basic information, it is difficult for many people to place Chiari in the spectrum of disease. Is it rare? Is it common? Does it affect a person's lifespan? Is it severe? Is it no big deal? Until these questions are answered scientifically, with quantifiable data, it will be challenging to raise awareness of Chiari and its impact.

Finally, the fact that Chiari is corrected surgically may play a role in the lack of research. Any disease which can potentially be treated medically with a drug, carries with it an inherent economic incentive. Millions of dollars are spent looking for neuropathic pain drugs (which is a side effect of Chiari) because the company which develops an effective one will make many times what they have spent on research. There is literally no economic incentive for a company to spend money researching Chiari.

A second hurdle to the possible surgical limitations of Chiari is the fact that neurosurgeons are a relatively small group who spend most of their time in surgery. A disease like MS attracts neuroscientists, neurologists, and other types of physicians and researchers, and studies are published in a range of medical journals. Conversely, there are very few Chiari publications outside of the neurosurgical arena.

While the road ahead to researching and understanding Chiari and syringomyelia appears to be a long one, it is important to remember that the longest journey starts with a single step. It is time for us, the patient community, to take that step by raising money for research and raising our voice to be heard.

--Rick Labuda

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