Special Report 01.08











Key Points

- There 161 English language studies published in 2007 involving Chiari and/or syringomyelia
- 44% of those were either Case Studies or mentioned Chiari incidentally
- 3. Surgical technique continues to evolve
- Researchers continue to explore different theories on Chiari

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, 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

Year In Research: 2007

Two thousand seven wasn't a bad year for Chiari research, but it wasn't a great year either. According to a PubMed search - PubMed is an online database of medical literature - there were 161 English language publications in 2007 (see Table 1) which at least mentioned Chiari and/or syringomyelia. While this is an increase from previous years, Chiari research continues to be dominated by Case Reports which simply describe one or two patients. Specifically 71 of the 161 citations (44%) were such Case Studies. Eliminating publications in which Chiari is incidental, reduces the number even further to 72 (see Table 2).

<u>Table 1</u> 2007 PubMed CM/SM Citations By Subject Type (161 Total)

Subject	# of Citations	
Total	161	
Incidental Reference	18	
Case Study	71	
Outcomes	8	
Surgical Technique	11	
Theories	8	
Diagnosis	2	
Symptoms	4	
Related Conditions	18	
Chiari II	3	
Animal	5	
CSF	2	
Morphological	4	
Reviews	2	
Scoliosis	4	
PTS	1	

- Incidental refers to a study which mentions Chiari or syringomyelia in passing, but CWSM 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

<u>Table 2</u> <u>Yearly CM/SM Research Citations (2004-2007)</u>

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

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

Although this represents and increase over previous years, the overall number is still very small compared to other diseases of a comparable nature. Plus, much of the increase from 2006 was due to publications focused on spina bifida.

Still, there is always good news to be found. In total there were 27 publications on surgical technique, outcomes, and theories about Chiari and syringomyelia. Also, morphological studies, which involve measurements of the craniocervical area, are becoming a widely accepted method of research into Chiari.

Some of the things we learned in 2007 include:

• Cerebellar tonsil tissue removed during surgery was clearly abnormal.

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 1/1/07 and 12/31/07. Duplicate results were eliminated manually. Citations were categorized by the editor. Foreign language publications were excluded.

- Adults with scoliosis tend to have poorer outcomes
- Bone only decompression is an emerging alternative children and possibly even adults
- Sleep apnea is extremely common among Chiari patients, but usually responds well to decompression surgery
- There are likely several underlying causes of Chiari, which can manifest with different symptomology and anatomical features
- Complex Chiari cases tend to have poorer outcomes

One of the highlights of the year was the UIC/Conquer Chiari Research Symposium. This one day event brought together more than 40 surgeons and scientists to present and discuss recent developments. The conference was such a success that Conquer Chiari is planning an expanded conference for 2008.

Outside of the Chiari arena, there was a major breakthrough in stem cell research. Scientists successfully used skin cells to create adult stem cells. The technique was replicated by several groups and appears to be fairly simple and stable.

Although there is still a long way to go for Chiari research to get to where it needs to be, in many ways 2007 was a much better year than 2006. Hopefully this represents the first small step up the mountain.

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