Key Points

1. The exact number of people with Chiari and/or syringomyelia is not known
2. Estimates have increased since the introduction of MRI technology, but still very few epidemiological studies
3. This study looked at the incidence and prevalence of SM in northern New Zealand
4. Found that the incidence of syringomyelia increased dramatically over time; especially after the introduction of MRI's in 1992
5. Found the overall prevalence of SM to be 8.2 per 100,000 in 2003
6. Found significant variations in the prevalence of SM between ethnic groups
7. Authors speculate there may be natural variations in posterior fossa size and/or CSF flow characteristics between ethnic groups
8. Also found that among Caucasians, Chiari related SM was more common in women than men

Definitions

epidemiology - the study of the distribution and causes of disease among people

incidence - the number of new cases of a disease in a year; expressed as the number of cases per a set number of people per year; for example 5 new cases per 100,000 people per year

Maori - the native, indigenous people of New Zealand

prevalence - the proportion of a given population that has a disease at a specific time; for example 10 cases per 100,000 people in the year 2006

retrospective - type of study which utilizes medical records and databases to look back at events that have already occurred

Syringomyelia In New Zealand

April 20, 2006 -- How many people have Chiari? How many people have syringomyelia? These seem like such simple questions, straightforward and easy to understand, yet more than 100 years after Chiari and syringomyelia were first identified they remain unanswered.

Whenever the mainstream media mentions Chiari and SM, they are always referred to as rare or very rare conditions. Indeed, even the National Institutes of Health, until recently, stated that less than 50,000 people in the US suffer from SM, and few people would hazard a guess on Chiari. However, with the rapid introduction and adoption of MRI technology, increased diagnoses have led to dramatically higher estimates for both.

Some people think 200,000 Americans have Chiari, some think more than half a million. Unfortunately, these are still just estimates. The truth is that there have been virtually no rigorous epidemiological studies on Chiari/syringomyelia, so we don't know for sure.

One way to determine how many people have a disease is to take a random sample of the population and see what percentage of the sample have it. If the sample is large enough, and truly random, the results can be extrapolated to the population as a whole. However, this approach is not practical when it comes to Chiari and syringomyelia. Taking thousands of people off the street, giving them MRI's and neurological exams, and analyzing the results would cost millions of dollars.

Another approach is to find a large group of people for whom medical records exist in one location. The records can then be analyzed to determine how many people suffer from a specific disease, and if possible, the results extrapolated to a larger population.

One such group is in northern New Zealand, specifically in Auckland and Northland. For these two regions, with a combined population of over 1 million people, there is only one hospital which provides neurosurgical services. Also, the government there funds the vast majority of healthcare, and keeps centralized records which are fairly uniform.

Recently, a group of researchers from the Auckland Hospital in New Zealand, led by Brickell and Anderson, took advantage of this situation to study the incidence and prevalence of syringomyelia in northern New Zealand. Specifically, they identified every case of syringomyelia diagnosed among residents there between 1961 - 2003. They published their results in March, 2006, in an on-line article in the Journal of Neurology, Neurosurgery, and Psychiatry.

In reviewing the hospital records, patients were considered to have SM if they had signs and symptoms consistent with syringomyelia and evidence of a syrinx on MRI, CT, myelograph, or during surgery. Patients with slit-like syrinxes, and no evidence that the syrinx was ever larger, were excluded, as were patients with a syrinx secondary to a spinal cord tumor, and non-residents.

Using this criteria, the team found 137 syringomyelia patients during the given time period. Perhaps not surprisingly, the majority (110) were diagnosed via MRI. As a group, the average age of diagnosis was 32 years, but symptoms first appeared on average at 27 years. In terms of men and women, the group was split almost evenly, and 64% of the cases were Chiari related.

When the researchers started analyzing the data, they found that the incidence (the number of new cases per year) increased dramatically during the time period studied (see Table 1). Between 1962-1971 there were only 0.76 new cases of syringomyelia per 100,000 people per year. By the 1980's this had increased to 2.15 new cases per 100,000 per year, but then really exploded when MRI technology was introduced in 1992. Between 1992 - 2001, the incidence increased to 4.7 new cases per 100,000 per year, more than 6 times the rate during the 1960's. Seeing this trend, it is understandable why syringomyelia was thought to be so rare.

In terms of prevalence - the proportion of people who have syringomyelia - the researchers calculated that in 2003, 8.2 people per 100,000 northern New Zealanders had syringomyelia. Surprisingly, they also found a significant difference among the major ethnic groups. The Pacific Islanders had the highest prevalence at 18.4 per 100,000, and the native Maori were also higher than average at 15.4 per 100,000. The difference between ethnic groups was especially pronounced for Chiari related syringomyelia.

The authors speculate that perhaps different ethnic groups naturally have different sized posterior fossas or CSF flow characteristics which result in more or less syringomyelia. It is interesting to note that in previous publications, Bogdanov, a Russian researcher, has identified an area in Russia with an unusually high number of syringomyelia cases. Based on these findings, future research into the average size and shape of the

spina bifida - birth defect where the spinal cord does not close properly; up to 30% of spina bifida patients also have Chiari

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

cerebellum - part of the brain located at the bottom of the skull, near the opening to the spinal area; important for muscle control, movement, and balance

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

decompression surgery - general term used for any of several surgical techniques employed to create more space around a Chiari malformation and to relieve compression

MRI - magnetic resonance imaging; large device which uses strong magnetic fields to produce images of soft tissue inside the human body

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

syrinx - fluid filled cyst in the spinal cord

Source

Brickell KL, Anderson NE, Charleston AJ, Hope JA, Bok AP, Barber PA. Ethnic differences in syringomyelia in New Zealand. J Neurol Neurosurg Psychiatry. 2006 Mar 20; [Epub ahead of print]

posterior fossa of different ethnic groups may provide additional insights.

While there were no significant gender differences in the group as a whole, there was one gender difference which stood out. Among Caucasians, Chiari related syringomyelia was much more common in women than men. This would seem to support the general notion, at least in the US, that Chiari affects more women than men.

So what do these prevalence numbers mean? While the overall prevalence finding seems low compared to recent estimates, it is important to note that the rapid increase in incidence rates, plus the variations between ethnic groups, makes applying these numbers beyond New Zealand difficult.

Based on this work, it is clear that any epidemiological study of Chiari and/or syringomyelia is very sensitive to the technology available to diagnose the conditions. It would not be surprising to find higher rates among groups of people for whom MRI is readily available or has been available longer. However, this research does raise some interesting questions regarding the possible differences among ethnic groups and their connection to the underlying causes of Chiari and syringomyelia.

Table 1
Incidence Of Syringomyelia Over Time

1962-1971	0.76
1972-1981	1.27
1982-1991	2.15
1992-2001	4.7

Note: Number represents the number of new cases per 100,000 people per year; MRI's were introduced to New Zealand in 1992.

Table 2
Prevalence Of Syringomyelia By Ethnic Group

Caucasian/Other	5.4
Maori	15.4
Pacific Islanders	18.4
Total	8.2

Note: Number represents the number of people with syringomyelia per 100,000 people in 2003.

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