

Key Points

- Previous research with small groups has identified a strong association between Chiari and sleep apnea
- Study out of France of 16 people with Chiari and SM found that 75% suffered from sleep apnea and 81% reported excessive daytime sleepiness
- 3. Almost half of the apnea was classified as central apnea
- 4. 8 of the 12 patients with apnea underwent decompression surgery, and 6 of these underwent additional sleep studies after their surgery
- 5. There was a significant decrease in the apnea index score after decompression surgery, especially central apnea
- Interestingly, there was NO significant change in the reports of daytime sleepiness
- Some subjects continued to suffer from severe obstructive apnea and required apnea related treatment to sleep
- Chiari is thought to cause apnea by either compressing the breathing control center in the brainstem; compressing the cranial nerves which carry important nerve signals, or by causing atrophy in the muscles of the lower throat

Definitions

apnea - temporary stop in breathing

brainstem - portion of the brain which connects with the spinal cord; controls many automatic functions such as heart rate, breathing, and swallowing

central sleep apnea - sleep apnea due to a delay in the nerve signal from the brain to breathe

cranial nerves - 12 pairs of nerves that start in the brain itself versus the spinal cord

hypopnea - slow or shallow

Decompression Surgery Helps Chiari Related Sleep Apnea

February 20, 2006 -- A recent study from France has provided more evidence that sleep apnea is a common, and significant, symptom associated with Chiari. Previous studies (see Related Articles) have identified a high rate of apnea among Chiari and syringomyelia patients. Now, a study led by Dr. Gagnadoux at the Centre Hospitalier Universitaire in France, and published in a recent addition of Neurology, found that 3 out of 4 patients with both Chiari and syringomyelia suffered from clinically diagnosed sleep apnea. They also found that decompression surgery significantly improved the Chiari related apnea.

Sleep apnea is a disorder characterized by repeated incidents where a person stops breathing, partially wakes up, then starts breathing again. The frequent episodes of apnea and arousal (also called microarousals) often lead to daytime exhaustion.

In general, there are three types of sleep apnea: obstructive, central, and mixed. Obstructive sleep apnea occurs because something physically blocks, or obstructs, the airway (muscles for example). In central sleep apnea, the problem lies with the respiratory control center itself, which for some reason fails to signal the body to breathe. Mixed refers to someone who suffers from episodes of both types of apnea.

In the French study, the researchers studied 16 people with both Chiari and syringomyelia - as demonstrated by MRI - to determine if one, they suffered from any type of sleep problems, and two, what effect, if any, decompression surgery would have on their sleep symptoms.

The participants completed a sleep questionnaire which evaluated items such usual length of sleep, apnea observed by partners, difficulty in waking up, chronic fatigue, morning headaches, waking during the night, snoring, and restless legs. They also completed the Epworth Sleepiness Scale (ESS), which measure daytime sleepiness.

To determine who was suffering from sleep apnea, each subject also underwent a full-night sleep study, or polysomnography. Polysomnography involves measuring brain waves during sleep and monitoring oxygen levels and respiration during the different sleep stages. When measured this way, apnea is defined as a stop in airflow of 10 seconds or more. An apnea episode is considered obstructive when there is an attempt to breathe during the episode and central when there is no attempt to breathe. Additionally, episodes of hypopnea, or a reduction in airflow of 30% or more can identified and tracked. The measurements and observations during a night of sleep testing can be combined into a single apnea/hypopnea index (AHI). Although definitions can vary, for this study, sleep apnea was defined as an AHI score greater than 10.

The results showed that on average the Chiari patients were not sleeping too well. From the questionnaires, more than 80% reported excessive daytime sleepiness and the average ESS score was 9.1, very close to the level (10) defined as being a problem. In fact, half of the group did have ESS scores above 10.

The polysomnography results were just as problematic, with an average AHI score of 36.6, and 12 out of the 16 subjects scoring above 10, meaning 75% of the group was diagnosed with sleep apnea. Some of the index scores were extremely high, with four people scoring above 70. Interestingly, nearly 50% of the apnea episodes were due to central apnea, where the breathing center in the brainstem fails to send the signals to breathe.

Next, eight of the twelve patients with sleep apnea underwent decompression surgery for their Chiari. Out of this group, six underwent a follow-up sleep study to see if the Chiari surgery helped their sleep apnea. The researchers found that the decompression surgery resulted in a decrease in the average AHI score from 56.5 to 37.5. While this was a significant decrease apnea problems still remained and there was no significant change in the average level of daytime sleepiness. This was likely due to the fact that a number of patients still had significant obstructive apnea after surgery. So, while the decompression significantly reduced the number of central apnea episodes, obstructive apnea remained a problem for some. In fact, two patients ended up using traditional apnea treatments (a device which produces positive airway pressure during sleep).

While the precise mechanism by which Chiari is linked to sleep apnea is not known, there are a number of possibilities. First, the Chiari malformation itself may compress the brainstem, which is where the breathing center is located. Second, Chiari is also known to compress and interfere with the function of the cranial nerves which are also important for breathing at night. Finally, Chiari is also known to cause problems in the throat area, such as swallowing, .htmly voice, etc. It may be that in some cases of Chiari, the muscles of the lower throat become weakened, and this weakness leads to an obstruction of the airway during sleep. While the authors of this study don't speculate, it might be that the decompression surgery was able to relieve pressure on the brainstem - leading to a decrease in central apnea, but the obstructive apnea remained a problem in some cases because throat muscles were already atrophied.

breathing

obstructive sleep apnea - sleep apnea due to an obstruction in the throat

polysomnography - studying physical measures - such as breathing - during sleep in a controlled environment

respiration - the act of breathing

sleep apnea - disruption of breathing during sleep which lasts longer than 10 seconds

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

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

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

Source

Gagnadoux F, Meslier N, Svab I, Menei P, Racineux JL. <u>Sleep-</u> <u>disordered breathing in patients</u> <u>with Chiari malformation:</u> <u>improvement after surgery.</u> Neurology. 2006 Jan 10;66(1):136-8. While several studies have now found a strong link between Chiari and sleep apnea, they all involved a relatively small number of people. Given the existing evidence, a larger scale study is now necessary to truly establish the incidence and characteristics of Chiari related sleep apnea. Given that extreme fatigue is a common symptom for people with Chiari and syringomyelia, one has to wonder if some of that fatigue is actually due to sleep apnea which could be treated.

<u>Table 1</u> <u>Changes In Sleepiness And Apnea Scores Before & After Surgery (6 Patients)</u>

Patient	ESS		AHI	
	Pre	Post	Pre	Post
1	9	14	26	6
2	12	10	20	10
3	12	18	73	82
4	9	10	88	75
5	0	0	54	18
6	16	12	78	22

Notes: ESS = Epworth Sleepiness Scale; AHI = apnea/hypopnea index; With both scales a score >10 is considered a problem; 8 out of 12 patients with apnea underwent surgery, but only 6 underwent sleep testing after surgery

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