

Key Points

1. Chiari and syringomyelia may cause compression of the brainstem area, plus interfere with nerve signals traveling to and from the brain
2. During sleep, breathing is controlled by the central respiratory system and relies on getting information from the body and sending signals to the body
3. As far back as 1941, there have been reports of Chiari interfering with breathing
4. There are case reports of sudden respiratory failure due to Chiari
5. After Chiari decompression, there is often a period of weakened breathing
6. True prevalence of sleep apnea in Chiari patients is not known, but limited studies have shown a very high rate compared to the general population
7. The exact mechanism by which Chiari/SM interferes with breathing is not yet known

Definitions

afferent - type of nerves which carry information from parts of the body back to the central nervous system

apnea - temporary stop in breathing

basilar invagination - condition, sometimes associated with Chiari, where the C2 vertebra is displaced upward, potentially compressing the brainstem

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

cerebellar tonsils - portion of the cerebellum located at the bottom,

Strong Link Between Chiari & Sleep Apnea

In a previous publication ([It Can Be Hard To Get A Good Night's Sleep With Chiari](#)), Dr. Botelho, a Brazilian sleep researcher, demonstrated that over 50% of Chiari patients suffered from some level of sleep apnea. Building on this primary research, Dr. Botelho and his colleagues recently published a review of the connection between Chiari and sleep apnea in the Journal Neurosurgical Review.

In the human respiratory system, the act of breathing is constantly monitored and adjusted in order to maintain the proper levels of oxygen, carbon dioxide, and pressure. To accomplish this, the central respiratory center (the part of the brain that controls breathing) receives signals from throughout the body which travel on afferent nerves. The respiratory center then processes this information and makes adjustments by sending commands to different parts of the body, such as the diaphragm.

An example of this system in action is when a person starts to exercise. Muscles work harder and send signals for more oxygen. The body responds, automatically, by increasing the breathing rate and adjusting whether the breathes are shallow or deep. All of this occurs in the part of the central respiratory system that controls involuntary breathing. This part of breathing central is located in the brainstem, just above the spine.

There is, however, a second part of the breathing control center which allows for voluntary control over breathing. Located in the cerebral cortex (which is responsible for higher order processes) this part of the brain takes over when a person consciously changes their breathing, for example to sing, or to take slow deep breaths in an effort to calm down.

This complex interaction between nerves carrying signals from the body, two parts of the brain deciding what to do, and nerves carrying commands back to the body, changes when a person goes to sleep. The voluntary breathing control center essentially shuts down. In addition, the involuntary center's response to stimulus (both inside and outside the body) is reduced. Finally, the muscles of the airway relax which results in an increase in resistance to the natural flow of air.

The changes in breathing during sleep are particularly evident during the REM stage of sleep (considered to be when a person dreams). During REM, breathing becomes very irregular and will switch quickly from rapid breaths to slow breaths and from shallow ones to deep ones.

During the altered breathing states of sleep, problems, which can be quite serious, sometimes develop. The term apnea refers to a temporary stop in breathing. 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 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.

While millions of people in the US who don't have Chiari suffer from sleep apnea, there is evidence that the rate of apnea in Chiari and syringomyelia patients is much higher than in the general population. According to Botelho's review, in 1941 a series of cases were published linking cranio-vertebral malformations to altered breathing. In addition, there have been a number of case reports of sudden respiratory distress linked with Chiari throughout the years.

In reference to sleep apnea tied specifically to Chiari, there again are a number of case reports, including ones where apnea is the first symptom of Chiari. While there are reports of apnea improving after decompression surgery, there are also indications that there is an increased risk of breathing problems in the period immediately after surgery (Ed. Note: I was given oxygen the first night after surgery, in the ICU, because I was having apnea type episodes while I slept).

Unfortunately, like so many other aspects of Chiari, the medical literature regarding Chiari and sleep apnea is dominated by case reports, so the true rate of sleep apnea among Chiari patients is not yet known. However, Botelho's own research group conducted two small studies which do indicate a high rate of sleep apnea.

Their first study involved analyzing 11 patients with Chiari, basilar invagination, and/or syringomyelia. Among this group, 72% were verified to have some type of apnea using sleep studies. In the team's second study, a group of 32 such patients were compared to a group of healthy volunteers. In the patient group, the overall rate of sleep/breathing problems was 59% compared to only 12% for the healthy group. Strikingly, 88% of the patients

so named because of their shape

cerebral cortex - part of the brain responsible for conscious experience, emotion, thought, and planning

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

efferent - type of nerves which carry signals, or commands, from the central nervous system to muscles and organs

hypopnea - slow or shallow 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

REM (rapid eye movement) - specific phase of sleep where dreams are thought to occur, characterized by rapid movement of the eyes

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

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

with basilar invagination were diagnosed with apnea problems. Interestingly, while central apnea was more common in the first study, the second study demonstrated a high rate of obstructive apnea as well.

While the exact mechanism linking Chiari and apnea has not been demonstrated, there are several likely candidates. Chiari often results in direct compression of the brainstem and can lead to a number of symptoms related to brainstem function, of which apnea may be one. In addition, Chiari often puts pressure on the cranial nerves (which originate in the brain as opposed to the spinal cord) which may result in atrophy of the muscles around the airway. This in turn could lead to an obstruction of the airway during sleep. Finally, the presence of a syrinx in the spinal cord may directly interfere with the important nerve signals which travel to and from the brain to monitor and control breathing.

As research into this very serious Chiari symptom progresses, it will be interesting to see if specific anatomical features are linked to it and whether, and how well, different surgical techniques help.

Figure 1 **Types Of Sleep Apnea**

- Obstructive Sleep Apnea - frequent episodes of partial or total blockage of the upper airway during sleep; often results in daytime exhaustion due to disturbed sleep
- Central Sleep Apnea - frequent episodes where someone stops breathing during sleep, but not due to blockage; often results in daytime exhaustion due to disturbed sleep; caused by interference with the central respiratory control center

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[Large Study Reveals Wide Range Of Chiari Symptoms](#)

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Source

Botelho RV, Bittencourt LR, Rotta JM, Tufik S. [Adult Chiari malformation and sleep apnea.](#) Neurosurg Rev. 2005 Jul;28(3):169-76

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