From The Desk of Regina S Bland, MD

I am a board certified pediatrician practicing in West Palm Beach, FL. I was diagnosed with Chari malformation in July, 1988 and have undergone a cervico-syringoarachnoid shunt in 1990 and decompression surgery in 2003. I take medications daily to enable me to be present for my patients. I hope that in this occassional article series that I might "bridge the gap" between patient and doctor.

Dr. Bland: Chiari & Sleep Apnea

January 20, 2006 - Sleep apnea (SA) is defined as the temporary absence or cessation of breathing (airflow) during sleep. It affects 2-4 % of the American population and its diagnosis has increased dramatically over the past few years. SA may lead to fragmented sleep with an associated decrease in daytime alertness and increased difficulty performing complex tasks. It may also be associated with an increase in cardiovascular risk and systemic hypertension. SA occurs in patients with Chiari 1 Malformation although the pathophysiology is unclear. A new study published in the Jan 10, 2006 issue of Neurology, the scientific journal of the American Academy of Neurology, shows that SA occurs in 75% of patients with Chiari 1 malformation and occipital decompression reduces the occurrences of SA by 90% in this patient population.

For the diagnosis of sleep apnea, airflow must be absent for some arbitrary period of time longer than the usual inter-breath interval. This is traditionally defined as 10 seconds for adults, and eight seconds (or more than two times the normal respiratory cycle time) for infants. Airflow can stop for various reasons:

In central apnea, no effort to breathe is made.

In obstructive apnea, there is ventilatory effort but no airflow because of closure of the upper airway.

In a typical mixed apnea, there is no initial ventilatory effort and an obstructive apnea pattern is observed when ventilatory effort is resumed.

The cause of apnea is not known but there are known risk factors. Obesity is a known risk factor and it is noted that the retro-pharyngeal airway is significantly smaller in this patient population. Upper airway obstruction (rhinitis, nasal polyps, deviated nasal septum, enlarged tonsils and adenoids) or facial malformations (micrognathia, etc) also are known to cause obstructive sleep apnea. Neurological and neuromuscular disorders may affect the strength or function of the upper airway or respiratory muscles or the respiratory control system.

Sleep apnea is diagnosed by some form of "sleep study" following clinical suspicion. These studies are often performed in "sleep laboratories" and involve measuring various parameters (usually EEG, EMG, airflow, airway pressure, PeCO2 (exhaled CO2) and PaO2 (aterial oxygen) levels). Indices have been developed that allow quantification of numbers of apnea in a specified time frame.

Once the diagnosis of sleep apnea is made, the etiology of the SA must be determined so that individual treatment can be provided . Weight control is the standard treatment in the obese. Medications are aimed at increasing the brain response to certain chemical measurements, eg Diamox (acetazolamide) has been used off-label to stimulate the respiratory center. Surgery may be aimed at relieving airway obstruction (eg. tonsillectomy and adenoidectomy, especially in the pediatric patient). And now, surgical treatment by occipital decompression has been shown to significantly decrease sleep apnea (especially central sleep apnea) in a group of Chiari 1 malformation patients.

In the current study, 12 of 16 patients with Chiari 1 malformation had SA. Of those, half were central SA. Eight of the 12 patients with SA had decompression surgery. Six patients with SA were studied post-operatively and were found to have 90% fewer occurrences.

The reason occipital decompression diminishes apnea is unclear. Occipital decompression may directly relieve pressure on the respiratory center located in the brain stem and may therefore reduce central SA. Occipital decompression may allow improved cranial nerve function (these individual cranial nerves innervate the muscles of the upper airway). It may allow normal nerve communications through the cerebellum (vital nerve bundles pass through the cerebellum and signals are repackaged and sent to appropriate brain centers).

It is important that research directed at Chiari 1 malformation (and its associated medical conditions) continues. It is important that these research studies are published in well-respected peer reviewed journals, such as Neurology, so that many more doctors are aware of the disease impact. It is important that this community disseminates this information to its members so that we can be active participants in our care.
