



## CPAP and BiPAP

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Airway obstruction while sleeping is commonly seen in individuals with MPS and related diseases. The neck is typically short and the airway can be unusually narrow. In addition, the tonsils and adenoids can become enlarged and block the airway, which can also contribute to breathing difficulties. A combination of these things cause obstructive sleep apnea, a common airway problem in individuals with MPS. Obstructive sleep apnea is defined as temporary breathing interruption during sleep because the airway in the neck becomes blocked as muscles in the airway relax. Sleep apnea can be improved in some individuals by opening the airway with nighttime CPAP (continuous positive airway pressure) or BiPAP (bi-level positive airway pressure).

Both of these treatment strategies are noninvasive. This type of breathing treatment does not involve surgery. It does require that the individual wear a mask and a mouthpiece while they sleep in order to receive the positive airway pressure to keep the airway open. While generally very similar, there are slight differences between CPAP and BiPAP. BiPAP differs from CPAP in that the pressure during expiration (breathing out) may be adjusted separately from the pressure delivered during inspiration (breathing in). CPAP and BiPAP treatments are very effective treatments for sleep apnea, but they do not cure the underlying problem.

CPAP means there is a continuous supply of air at the same pressure being delivered to the patient with each breath. Nasal CPAP delivers air pressure through the nose. The mask is placed securely over the patient's nose and slight positive air pressure is used to increase the amount of air being inhaled without making the work of breathing more difficult. The mask does not breathe for the patient. The airflow creates enough pressure when inhaled to keep the airway open. Another method for delivering air to a patient involves placing a tube into the nose, in order to supply gentle air pressure to the airway.

Occasionally, CPAP can increase the work associated with breathing. In those cases, BiPAP is prescribed instead. BiPAP is also referred as non-invasive face mask ventilation. Bi-level means that with each breath the individual takes, the pressure changes. The pressure rises when inhaling and drops when exhaling, making breathing easier. BiPAP therapy is usually prescribed for patients with sleep apnea if the CPAP therapy is too difficult. Occasionally the BiPAP machine will be spontaneously timed (BiPAP SP); meaning that if for some reason the patient does not take a breath, the machine will automatically initiate a breath for them.

The major obstacle that most people must overcome is adjusting to the CPAP or BiPAP system. These patients must get used to sleeping while wearing the mask and mouthpiece. Approximately 20% of individuals never adjust or grow accustomed to the treatment method. Some people feel that the device is claustrophobic. Others find that it is difficult to transport with them when traveling.

Some complications can arise through the use of positive airway pressure devices. These complications can be associated with the equipment or with the patient's clinical condition. Mucus can build up in the nasal tubes. Patients can also become uncomfortable if the pressure is set inappropriately high.

To determine whether someone has sleep apnea and may benefit from CPAP or BiPAP, a doctor can arrange for a sleep study. During a sleep study, a person is continually monitored for apnea episodes or decreases (desaturations) in their oxygen level. An abnormal sleep study may suggest that CPAP or BiPAP would be helpful to maintain an open airway during the night. Before considering any therapy or treatment, consult your pulmonologist.

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