Use and misuse of CPAP in neuromuscular disorders

Hypoventilation occurs when a reduced amount of air enters the lungs resulting in decreased levels of oxygen and increased levels of carbon dioxide in the blood . When Hypercapnia(the abnormal retention of carbon dioxide (PCO2) is detected noninvasive positive pressure ventilation (NPPV) is offered to support the weaker respiratory muscles during sleep. There is a wealth of evidence that this technique improves nocturnal ventilation and sleep quality

Continuous positive airway pressure (CPAP) is the typical recommended treatment for obstructive sleep apneas (OSA). Sometimes, OSA can be present in Myotonic dystrophies or in other NMDs.

CPAP delivers constant air pressures during the breathing cycles ,thereby operating merely as a splint to keep the airways open . As It delivers constant positive inspiratory and expiratory pressures, It does not assist the weakened inspiratory muscle function and is unable to deliver sufficient volumes of air to increase inspiratory intake. The resulting reduced oxygen intake and retention of CO2 may enhance hypoventilation increasing morbidity and mortality and decrease quality of life in those patients with ventilatory pump failure.

The use of CPAP presupposes that respiration is fully supported by normally functioning respiratory muscles**. This is precisely where the problem lies.**

Patients affected by NMDs have weak skeletal and respiratory muscles. Therefore the recommended treatment to improve hypoventilation is NPPV *(can we say BIPAP)* rather than CPAP. In Bilevel positive airway pressure ventilation, typically called BiPAP, or VPAP. Higher pressure air is delivered on inhalation and a lower (or zero) pressure on exhalation. In muscle diseases, the difference between the inhalation and exhalation airflow pressures, is typically quite high to provide greater assistance to the inspiratory muscles and little or no resistance during exhalation.

Because sleep/breathing problems can a troublesome symptom for many people living with neuromuscular conditions, it is important **t**o consult an experienced, sleep specialist who is familiar with neuromuscular conditions and is able to advise on which therapeutic solutions are appropriate.

The problem arises when physicians mistakenly think that the problems are a result of central and obstructive apneas, and hypopneas, This results in the sleep specialists suggesting CPAP.

This is not a good idea since this therapy is dangerous and inadequate in patients with NMDs. In addition, given inevitable respiratory decline in NMDs, initiation with NPPV (BiPAP) rather than CPAP may avoid the need for device switching later.

In conclusion, we strongly suggest that CPAP should not be offered to individuals with neuromuscular disorders, as it is unable to treat nocturnal hypoventilation due to respiratory muscle weakness.

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