Sleep Related Breathing Disorders in Neuro-Degenerative Disorders and Stroke

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Management of sleep disordered breathing in neurological diseases: organizational and economical aspects
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In 2004 a task force to develop guidelines for diagnostic evaluation and treatment of sleep disorders in degenerative neurological disorders and stroke was initiated by the European Federation of Neurological Societies (EFNS). The aims of the task force were to provide evidence-based recommendations and to highlight the role of the neurologist in the management of sleep disorders associated with degenerative neurological disorders and stroke. Patients with neurological diseases often have significant sleep disorders including sleep related breathing disorders including obstructive sleep apnoea syndrome (OSAS), obstructive sleep apnoea syndromes (OSAS), central sleep apnoea-hypopnoea syndrome (CSAHS), Cheyne-Stokes Breathing Syndrome (CSBS) and sleep related hypoventilation/hypoxemic syndromes (SHVS), sleep fragmentation insomnia, sleep related motor disorders and REM behavioural disorders which may affect both nocturnal sleep and day-time function with increased morbidity and even mortality. A polysomnography is usually a diagnostic minimum for the diagnoses of the most commonly reported sleep disorders in patients with neurological diseases, eventually supplied with a full video-PSG/video-EEG-PSG should be considered in patients with nocturnal motor and/behaviour manifestations. Respiratory polygraphy has a moderate sensitivity and specificity in the diagnosis of OSAS without neurological diseases, but its value for diagnosis of SBD in patients with neurological diseases has not been evaluated as compared to gold standard PSG. Oximetry has a poor-moderate sensitivity-specificity for the identification of OSAS in patients without neurological diseases. CPAP is the most effective treatment of OSAS. This probably also includes patients with OSAS and neurological diseases. Bi-level PAP/variable PAP and volumetric ventilation are useful for SBD like CSAHS, CSBS and SHVS, especially in neuromuscular diseases. There is a need for further studies focusing on the diagnostic procedures and treatment modalities in patients with sleep disorders and degenerative neurological diseases and stroke.

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Sleep-related disordered breathing and stroke
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More than half of stroke patients (pts) exhibit SDB. Obstructive sleep apnea (OSA) is most commonly found. Central sleep apnea (CSA) and/or Cheyne-Stokes breathing (CSB) are seen particularly early after stroke onset and/or in patients with cardiac insufficiency. SDB usually improves after the acute phase. Post-stroke SDB can be a consequence of brain damage per se (large/strategically located lesions, lesions interfering with upper airway patency, stroke complications such as aspiration, immobility, respiratory infection). Recent studies suggest that SDB represents also an independent risk factor for stroke. Mechanisms involved in this link may include arterial hypertension, atrial fibrillation, reduced glucose tolerance, changes in cerebral hemodynamics, hypercoagulability and increased vascular oxidative stress/inflammation secondary to recurrent arousals and hypoxias observed with SDB. The presence of SDB in stroke pts is of clinical relevance having been associated with early neurological worsening, higher blood pressure values/variability in the acute phase, longer hospitalisation, and poorer long-term outcome (and rehabilitation potentials). SDB should be particularly suspected in male patients over the age of 40, with history of frequent snoring, diabetes, hypertension and night-time onset of stroke. SDB can be diagnosed by simple bedside tests (respirography, automatic CPAP devices). Compliance to CPAP treatment is low (about 50% in the acute, 20% in the chronic phase). In stroke patients with CSA/CSB improvement can be achieved with oxygen.

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Management of stridor and other sleep-related disordered breathing disorders in movement disorders
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Sleep-related disordered breathing (SDB) occurs in patients with movement disorders, particularly in multiple system atrophy (MSA) and Parkinson’s disease (PD) and may be associated with sleepiness, fatigue or reduced survival. SDB probably appears in at least 10–20% of movement disorder patients. In PD there are abnormal airflow oscillations and upper airway obstruction during wakefulness, probably related to deficient glottic and supraglottic motor function. Whether these changes persist during sleep is unknown. Sleep apnea (SA) has been reported in 13–66% of PD patients, more often than in controls and more frequent in sleepy than in non-sleepy PD patients, although these findings have not been found by all authors. SA in PD improves with continuous positive airway pressure (CPAP). Several breathing problems may occur during wakefulness and sleep in MSA, but the most relevant is stridor. Stridor occurs in 13–42% of patients, unrelated with the subtype (cerebellar or parkinsonian), is a marker of short survival and presents normally a few years after disease onset, although may also be its first manifestation. Stridor usually appears during sleep and after a variable interval may also occur during wakefulness. Both dystonia of the vocal cord closing muscles and paresia of the opening muscles have been held responsible for stridor. CPAP during sleep resolves stridor and is generally well tolerated by patients and bepartners. There is not enough evidence to support botulin toxin in the treatment of nocturnal stridor. When stridor becomes diurnal CPAP is no longer useful and tracheostomy should be considered.

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Sleep-related breathing disorders in neuromuscular diseases
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Neuromuscular patients are at high risk for the development of sleep related respiratory disorders and respiratory failure. Diaphragm weakness or paralysis is the most important determinant, but consequences of chest-wall deformity and weakness of the pharyngeal
muscles can also play a role. In a neuromuscular clinic where patients are regularly followed almost 50% will present with sleep and breathing abnormalities. The normal sleep-related changes in respiration put the neuromuscular patient at a specific ventilatory risk with development of nocturnal and then diurnal hypoventilation. During non-REM sleep there is an overall reduction in ventilation that is related to sleep state changes and the chemical control of breathing. During REM sleep there is a loss of tone in rib cage and other respiratory accessory muscles, leaving the diaphragm alone to maintain ventilation. Any process affecting the diaphragm (innervation or myopathy) will cause significant changes in breathing and oxygenation. Depending on the type of neuromuscular disorder, breathing abnormalities during sleep include prolonged periods of hypoventilation and apneas (central and/or obstructive), however obstructive sleep apnea syndrome does not occur more frequently than in the general population except for diseases with bulbar or pharyngeal involvement. Sleep breathing disorders contribute to frequent arousals, sleep time reduction and sleep deprivation. These abnormalities will progressively change ventilatory and arousal responses to changes in oxygen and CO2 levels. To compensate for muscle weakness or paralysis, respiratory centers become more tolerant to higher level of CO2 and O2 desaturation. Hypoventilation begins during REM sleep, followed by non-REM sleep and finally becomes permanent (awake and sleep). Symptoms are frequently insidious, not always well perceived by patients and are frequently only reported after their resolution by nocturnal ventilation. Symptoms include poor quality of sleep, morning headaches, daytime sleepiness and fatigue. Additional symptoms must be considered as possibly related to hypoventilation, such as weight loss, increase in swallowing disorders and recurrent respiratory infections. Conventional polysomnography with O2 saturation and ideally transcutaneous CO2 allow to distinguish among the different causes of sleep disturbance and to assess the severity of the disorder. Polysomnography is not easy for neuromuscular patients due to paralysis, poor comfort and pain related to orthopaedic problems. Interpretation of respiratory events should be carefully done as confusion between central and obstructive can occur due to muscle weakness. Polysomnography should be performed in all symptomatic patients, or when a significant reduction of vital capacity (< 50% of theoretical) is found, or systematically. Nocturnal hypoventilation should lead to the implementation of non invasive ventilation (NIV) as O2 or CPAP will not correct hypoventilation and risk to aggravate this condition. NIV will correct hypoventilation and restore better quality of sleep in spite of the inconvenience of the equipment (mask and ventilator and persistence of other causes potentially disturbing for sleep). Long-term, especially for slowly progressive disease, NIV will prolong survival, comfort and quality of life.

**Conclusion:** Sleep is a state of vulnerability for neuromuscular patients and polysomnography should be part of their routine respiratory evaluation to initiate appropriate treatment.