0802

CENTRAL SLEEP APNEA AS A RESULT OF CEREBRAL CAVERNOUS MALFORMATION HEMORRHAGE IN THE PEDIATRIC POPULATION

Joshua Bowling[1], Fauziya Hassan[2]
University of Michigan Sleep Disorders Center [1] University of Michigan Sleep Disorders Center, Division of Pediatric Pulmonology, Department of Pediatrics [2]

Introduction: Cerebral Cavernous malformations (CCMs) consist of a collection of capillaries in the central nervous system (CNS) that are enlarged and irregular in structure. Patients with CCMs are at increased risk of hemorrhage into the brain or spinal cord, resulting in seizures, focal neurologic deficits, hydrocephalus, and death. Given the importance of the brainstem and central chemoreceptors in regulating respiratory function, rupture of CCMs can also lead to dysregulation of breathing. We present a series of 3 pediatric patients who have a diagnosis of central sleep apnea (CSA) due to rupture of CCMs managed with home ventilators with significant differences in clinical presentation from mild CSA to night time respiratory support for severe CSA and chronic respiratory failure requiring continuous mechanical ventilation.
Report of Cases: Subject 1 is a 13 yo M with a history of ruptured posterior fossa arteriovenous malformation (AVM) complicated by post-hemorrhagic hydrocephalus, right hemiparesis and severe central apnea requiring nighttime mechanical ventilation via tracheostomy. Subject 2 is a 19 yo M with a history of mid-pontine cavernoma with multiple episodes of hemorrhage failing surgical resection resulting in right hemiparesis, severe central apnea and hypoventilation requiring continuous mechanical ventilator support via tracheostomy. Subject 3 is a 6 yo F with a history of thoracic cavernous hemangioma resulting in spinal cord injury and mild central sleep apnea requiring mask ventilation via home ventilator during sleep. All of these patients experienced loss of respiratory drive as a result of complications from brainstem or spinal cord lesions with varying degrees of ventilator support requirement and clinical presentation.
Conclusion: Due to the propensity for CNS bleeds among patients with CCM they are at increased risk of respiratory compromise. Home ventilator support can be used effectively to treat central apneas and chronic respiratory failure but this is a moving paradigm as subsequent bleeds worsen respiratory compromise.
Support (If Any): Support (if any):

0803

EXTREME UPPER EXTREMITY MOVEMENTS IN PATIENT WITH NARCOLEPSY TYPE 1 AND REM SLEEP BEHAVIOR DISORDER AFFECTING ACTIGRAPHY RESULTS.

Jack Galagan[1], Carla York[1], Rodolfo Soca[1]
Sleep Disorders Center, Walter Reed National Military Medical Center [1]

Introduction: Narcolepsy with Cataplexy is a central disorder of hypersomnia that is characterized by excessive daytime sleepiness and Rapid eye movement (REM) dissociation phenomena. A common manifestation of narcolepsy is REM behavioral disorder (RBD), a parasomnia characterized by loss of muscle atonia during REM sleep (RSWA). While RBD is relatively common in patients with narcolepsy, very extreme movements are considered rare.
Report of Cases: A 20-year-old male with no significant past medical history presented with 8 months of new onset daytime sleepiness, sleep paralysis, and auditory hallucinations at sleep onset. After initial evaluation at our sleep center, he was scheduled for actigraphy testing and sleep logs, followed by video polysomnogram and mean sleep latency testing (MSLT). Initial PSG results were notable for a total sleep time of 498 minutes, a normal apnea-hypopnea index of 0.7/hr., sleep onset latency of 0 minutes, and REM latency of 0 minutes. Based upon military medical standards, the MSLT scheduled for the following morning was cancelled due to limited sleep during the preceding 2 weeks as measured via actigraphy. However, the patient's sleep logs reported over 7.5 hours of sleep per night. The patient's video PSG was reviewed, which showed evidence of frank RSWA, as well as episodes of dream enactment behavior during REM sleep, all including very violent movement in his upper extremities. Repeat trial of MSLT 2 weeks later showed mean sleep latency of 1.7 minutes, with 5 sleep onset REM periods (SOREMPS), and evidence of dream enactment involving the upper extremities during 3 of these SOREMPS.
Conclusion: We present the case of a patient with narcolepsy and RBD with significant upper extremity movements to the point of affecting the actigraphy sleep/wake detection algorithm. Actigraphy has been validated as a diagnostic tool in assessing sleep and wake patterns in individuals without significant REM sleep dissociation phenomena; however, our case highlights the necessity of further research of the validity of actigraphy in patients with Narcolepsy and/or REM behavioral disorder.
Support (If Any):

0804

ENHANCED DRUG-INDUCED SLEEP ENDOSCOPY: DISTINGUISHING CENTRAL FROM OBSTRUCTIVE APNEAS

Crystal Cheong[1], Alan Schwartz[2], Everett Seay[2], Jorge Mora[1], Erica Thaler[2], Raj Dedhia[2]
Division of Sleep Medicine, Department of Medicine, University of Pennsylvania [1] Perelman School of Medicine, Department of Otorhinolaryngology–Head and Neck Surgery, University of Pennsylvania [2]

Introduction: Drug-induced sleep endoscopy (DISE) is a useful tool for assessing upper airway collapse in patients with obstructive sleep apnea (OSA) and frequently influences surgical plans. The standard DISE setup of an endoscopic tower with flexible bronchoscope is adequate for visualizing collapse configurations, but endoscopic findings do not always correlate with actual respiratory physiology. We describe our enhanced clinical DISE setup incorporating nasal flow and respiratory effort measurements, which facilitates differentiation between central and obstructive events. Central sleep apnea was detected in two patients who were originally diagnosed with OSA and underwent DISE during hypoglossal nerve stimulation candidacy workup.
Report of Cases: Case 1 is a 58-year-old male with cardiomyopathy, atrial fibrillation and congestive heart failure who was diagnosed with moderate OSA on a home sleep apnea test. He was PAP intolerant due to claustrophobia. DISE during baseline breathing revealed complete anteroposterior collapse at the palate, tongue base and epiglottis. However, central apneas with Cheyne-Stokes breathing were noticed when positive airway pressure (PAP) was applied. A subsequent polysomnogram revealed severe sleep apnea which was primarily central in nature with Cheyne-Stokes breathing and OSA. He declined retrying PAP and opted

for phrenic nerve stimulation. Case 2 is a 48-year-old male with a history of aortic valve repair, hypothyroidism, atrial fibrillation provoked by excessive thyroxine, hypertension and moderate OSA diagnosed on several polysomnograms. He had been treated previously with nasal and palatal surgery, oral appliances and PAP. DISE revealed complete anteroposterior collapse of the palate and tongue base, but also central apneas during baseline breathing and at low PAP levels. Polysomnogram performed following his DISE confirmed central sleep apnea which was positional in nature. He chose to undergo positional therapy instead of PAP or phrenic nerve stimulation.

Conclusion: The nasal flow and respiratory effort measurements included in our enhanced DISE setup enable the sleep surgeon to recognize the absence of respiratory effort even in the face of soft tissue collapse observed on videoendoscopy. These cases demonstrate the ability of propofol to preserve pathophysiologic mechanisms of sleep apnea (i.e. central versus obstructive), and underscore the importance of DISE as a diagnostic tool prior to sleep surgery.

Support (If Any): NIH 1R01HL144859-01

0805

HALO-TRACTION INDUCED OBSTRUCTIVE SLEEP APNEA

Vignesh Nayak[1], Kyle Bliton[1], Mary Maddox[1]
University of Alabama at Birmingham [1]

Introduction: Obstructive sleep apnea (OSA) is the most common sleep-related breathing disorder. It is a multi-factorial disease with a variety of identified causes including age, male gender, obesity, craniofacial and upper airway abnormalities. We would like to describe a patient who had severe OSA following application of Halo traction, which significantly improved following the removal of the device.

Report of Cases: 14-year-old male with medical history of spina bifida, chiari malformation s/p decompression, shunted hydrocephalus and severe scoliosis, was admitted to the hospital for anterior spinal discectomy L2-S1 and Halo application with traction for scoliosis. He previously had nocturnal polysomnogram (NPSG) in 2017 that demonstrated very mild mixed apnea with an apnea hypopnea index (AHI) of 5.5. Because central apneas were very brief and clustered in REM, family elected to repeat a study rather than treat. In 2019, he had a follow up study with complaints of snoring and thirst, and this demonstrated an AHI of 21 with 29 brief central apneas and 72 hypopneas, 1 obstructive apnea. He had a T&A and turbinate ablation and due to the global pandemic did not undergo repeat sleep study. During admission for his anterior spinal discectomy and Halo, he demonstrated persistent night time hypoxia. A split night sleep study showed evidence of severe OSA with pretreatment AHI of 94.4, oxygen nadir 86%. Continuous positive airway pressure (CPAP) was initiated at 5 cm of water and titrated to 11 cm of water. On CPAP of +11 severe obstructive events continued with an AHI of 40.6, oxygen nadir 92%. A bilevel positive airway pressure (BIPAP) titration study the subsequent night started at pressures of 12/6 and titrated to 21/9 with respiratory rate of 12 yet demonstrated AHI of 51, oxygen nadir 89%. Study transitioned to average volume assisted pressure support (AVAPS) with IPAP max of 26, IPAP minimum of 12 EPAP of 9, tidal volume of 175ml, rate of 12 with inadequate control of his obstructive events with an AHI of 24.8, minimum oxygen saturations of 91. While hospitalized, he remained on AVAPS with normal capillary blood gases. Halo traction was removed 2 weeks following his surgery with plan was to send him home on AVAPS and repeat NPSG in 6 weeks. However, as a result of COVID pandemic/Philips recall, CPAP was the only device available for home use, so CPAP therapy at +8 cm was trialed overnight, demonstrating oxygen nadir of 92% and a normal capillary blood gas in the morning. Patient was then discharged home on CPAP of +8 cm of water. He returned back to sleep center for a BIPAP titration study to re-establish BIPAP/AVAPS settings, as his inpatient sleep study had shown severe OSA. During the sleep study, he was started on BIPAP 12/6 and he remained on it throughout the night with 0 central and 0 obstructive events. As he did well, he was advised to continue CPAP +8 with plans to repeat the sleep study off CPAP. In clinic follow up, he reported mild skin breakdown and occasionally waking unrefreshed.

Conclusion: As our patient did significantly better following the removal of Halo traction device, it is likely that Halo traction device caused fixed over flexion of the cervical spine that resulted in decrease in his airway diameter, which further worsened during his sleep, and caused severe OSA.

Support (If Any):

0806

REM SLEEP WITHOUT ATONIA IN THE SETTING OF DUCHENNE MUSCULAR DYSTROPHY: CAN IT SERVE A PROTECTIVE ROLE?

Kayvon Sarrami[1], Jay Patel[1], Mahtab Moshtagh Sisan[1], Alon Avidan[1]
University of California, Los Angeles [1]

Introduction: Duchenne muscular dystrophy (DMD) has well-known associations with sleep-related breathing disorders such as sleep apnea and nocturnal hypoventilation. These disorders naturally follow from the progressive muscle weakness that is the hallmark of DMD. New data is emerging on the prevalence of other polysomnographic features affecting patients with DMD, particularly as they relate to the phenotype of the sleep disorders unique to DMD. We report on a patient with REM sleep without atonia (RWA) in DMD.

Report of Cases: A 13-year-old male with DMD was referred for a polysomnogram (PSG) due to new-onset snoring, but there was no history to suggest dream enactment behavior. He was not taking any CNS-acting medication such as serotonergic or sedative-hypnotic medications. Neurologic examination was notable for diffuse symmetric muscle weakness requiring the use of a wheelchair. PSG revealed obstructive sleep apnea syndrome with an apnea-hypopnea index of 3/hr overall and 5/hr during REM sleep, a respiratory disturbance index of 9/hr with minimum O saturation of 84%, and evidence of RWA in three 30-second epochs meeting AASM scoring criteria. Dream-enactment behavior was absent during the PSG and historically.

Conclusion: We report on a previously unrecognized finding of RWA the setting of DMD. While sleep studies are routinely performed in DMD due to the significant association with sleep-disordered breathing and REM-related hypoventilation, the presence of RWA was unusual. The finding of RWA in the setting of a dystrophinopathy prompts important questions. While it is unlikely that RWA is an emerging sign of an α-synucleinopathy in the setting of DMD, its presence in a young teenager raises the possibility of hypocretin deficiency in the setting of DMD, as is the case with myotonic dystrophy type 1 reflecting impaired hypocretin neurotransmission. However, a more attractive explanation is that RWA may promote a protective effect against sleep apnea in patients with vulnerability to REM-related hypoventilation. Our data suggest a potential protective role conferred by the preservation of muscle tone in the setting of sleep apnea in neuromuscular conditions.

Support (If Any): None