

O₂ at 3-4 L/min. Per the parents, the patient has been maintaining her oxygen saturation in the absence of BIPAP therapy with oxygen use. Due to COVID, patient was unable to follow up but will be scheduled for a repeat PSG in the near future. She followed with Neurosurgery for Arnold Chiari II and they recommended no surgical intervention at this time due to functional VP shunt.

Conclusion: This is an atypical presentation of Biot's breathing in the absence of CNS infections and opioid use in a patient with Arnold Chiari malformation II. Patient has complex sleep apnea, initially well controlled with BiPAP ST, but developed BiPAP intolerance. She is on oxygen with good control of hypoxemia in the absence of BiPAP therapy.

Support (If Any):

0837

AVERAGE VOLUME-ASSURED PRESSURE SUPPORT (AVAPS) AFTER CPAP FAILURE IN A PEDIATRIC PATIENT WITH SEVERE OBSTRUCTIVE SLEEP APNEA AND SLEEP-RELATED HYPOVENTILATION

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Introduction: Obstructive sleep apnea (OSA) has become an increasingly pervasive sleep disorder in the pediatric population. Current mainstream treatments include adenotonsillectomy and positive airway pressure therapy. Average volume-assured pressure support (AVAPS) is a relatively new mode of non-invasive ventilation, which has been increasingly used in the treatment of respiratory failure and hypoventilation syndromes. Here we present a case of a pediatric patient with severe OSA and sleep-related hypoventilation who was successfully treated with AVAPS after failure of CPAP therapy.

Report of Cases: A four year old boy with history of severe OSA, severe obesity, asthma, and allergic rhinitis underwent polysomnography one year after adenotonsillectomy and nasal turbinate reduction due to continued symptoms of sleep-disordered breathing. Results showed elevated residual apnea-hypopnea index (AHI = 30.4 events/hour), sleep-related hypoventilation (T ETCO₂ ≥ 50 = 228.3 minutes), and sleep-related hypoxemia (T ≤ 90% = 7 minutes). Therefore the patient underwent repeated adenotonsillectomy and turbinate reduction, with post-operative course complicated by pulmonary edema requiring intubation. He was extubated and weaned to nocturnal CPAP. Following discharge, CPAP titration failed to control AHI at maximal pressure (AHI 54.5 on 20 cm H₂O, T ≤ 90% = 15.3 minutes). The patient was then started on AVAPS with auto-titrating EPAP (AVAPS-AE, settings Pmax 20 cm H₂O, PS 2-10 cm H₂O, EPAP 5-10 cm H₂O, RR auto, room air) with subsequent improvement of snoring and witnessed apneas, as well as reduction of daytime sleepiness. Afterwards, AVAPS-AE titration confirmed resolution of obstructive sleep apnea, sleep-related hypoxemia, and sleep-related hypoventilation (AHI = 2.5, T ≤ 90% = 1.2 minutes, T ETCO₂ ≥ 50 = 6.5 minutes.) The patient has since remained stable on AVAPS-AE until age ten, with the most recent AVAPS titration demonstrating continued resolution of sleep-disordered breathing.

Conclusion: AVAPS was an effective treatment for a pediatric patient with severe OSA and sleep-related hypoventilation who had failed CPAP therapy.

Support (If Any): None.

0838

CHEYNE-STOKES BREATHING IN A PEDIATRIC PATIENT WITH DILATED CARDIOMYOPATHY AND MUSCULAR DYSTROPHY PRIOR TO HEART TRANSPLANT

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Introduction: Cheyne-Stokes breathing (CSB) has rarely been identified in the pediatric population. Neuromuscular diseases (NMD) such as Duchene Muscular Dystrophy (DMD) can predispose patients to sleep-disordered breathing including central sleep apnea (CSA) and CSB. Sleep-disordered breathing in children with NMD may not have symptoms; thus, treatment can be delayed. Currently, there is limited data to support resolution of CSB in DMD with dilated cardiomyopathy post-transplant.

Report of Cases: We present a 15-year old female with a significant history of both dilated cardiomyopathy and DMD who presented with acute on chronic heart failure. Due to her disease progression, she was listed for heart transplant. Prior to her transplant, she completed an inpatient polysomnography (PSG) to rule out sleep-disordered breathing due to concerns of snoring and dyspnea during sleep. Her Pediatric Daytime Sleepiness Scale score (PDSS) was 8. The polysomnogram recorded moderate obstructive sleep apnea (OSA) and central sleep apnea (CSA) consistent with Cheyne-Stokes breathing along with rare premature ventricular contractions (PVCs). Patient was started on BPAP of 13/8 cm H₂O with a back-up rate of 12 breaths per minute after titration study. The patient subsequently received a heart transplant in which the patient's dyspnea and snoring resolved. Post-transplant PSG pending to reassess the severity of sleep-disordered breathing.

Conclusion: Though CSA can be seen in children, CSB is rarely seen in children with either heart failure or muscular dystrophy. When CSB is observed, the cornerstone of treatment is correcting the underlying cause. This patient demonstrated CSB with symptoms that improved with BPAP and now post-heart transplant. When both heart failure and neuromuscular disease are involved, close monitoring for clinical symptoms along with screening for CSB is important and may affect overall quality of life and recovery.

Support (If Any):

0839

COMPLEX SLEEP APNEA IMPROVED WITH DECOMPRESSION OF A CHIARI I MALFORMATION IN A PEDIATRIC PATIENT

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Introduction: Chiari malformation (CM) occurs when a portion of the cerebellum herniates through the foramen magnum. CM is categorized as two types. Type 1 involves the cerebellar tonsils and type 2 involves the cerebellum and brain stem. Those with CM can be asymptomatic to having debilitating neurologic symptoms such as dysphagia, tinnitus, emesis, balance difficulty, muscle weakness, and/or headache. Central sleep apnea (CSA) and obstructive sleep apnea (OSA) have been associated with CM. It is postulated that