0530

A STUDY OF PEDIATRIC PAP COMPLIANCE IN LONGITUDINAL AND SINGLE-VISIT PATIENTS

Marilyn Culp[1], Shan Luong[1], Emily Nguyen[1], Anna Wani[1]
Children's Health/UT Southwestern Medical Center [1]

Introduction: PAP compliance is difficult to attain and maintain, but important for treatment efficacy in obstructive sleep apnea. This study sought to identify patterns of compliance by age, stratified by gender and device type in patients who followed up only once after treatment initiation (single-visit patients) and those who were seen more than once (longitudinal patients).
Methods: Charts for approximately 10 months from 10/2018 to 7/2019 were retrospectively reviewed for 1177 patient visits, representing 521 patients with 246 single-visit and 275 longitudinal patients. Some longitudinal patients had visits dating back to 2002. Data analyzed included: age at visit versus compliance measured by percentage of days used and percentage of days used over 4 hours during their compliance period. Data was analyzed for single visit and longitudinal patients stratified by gender and device type. Age was categorized as 0 to 24 months, then years 3.0 to 5.9, 6.0 to 12.9, 13.0 to 18.9 and 19.0 to 21.9.
Results: Patient demographics: 47.2% single-visit, 52.8% longitudinal; 60% male; Approximately 80% between 6 and 18.9-years-old; 80.8% used CPAP. Statistically significant results:Seen for different age groups among females (p=0.012): Females 19 to 21.9-years-old had 45.8% lower compliance for percentage of days used over 4 hours compared to 3 to 18.9-years-old.Longitudinal patients showed 7.5% and 2.4% better compliance for percentage of days used (p=0.0006) and for days used over 4 hours (p=0.002), respectively, compared to single-visit patients after adjusting for sex and age group. Using a BPAP device was associated with 8.9% better compliance for use over 4 hours (p=0.002) compared to CPAP after adjusting for sex, age and visit groups. Longitudinal patients had 7.7% higher compliance for % days used over 4 hours compared to single visit patients (p=0.0016) after adjusting for sex, age group and device.
Conclusion: Statistically significant positive factors for compliance were longitudinal patients versus single-visit patients and those using BPAP.Among females, 19 to 21.9-year-olds were least compliant, otherwise no differences between age or gender were identified.
Support (If Any):

0531

CHARACTERIZATION OF SLEEP-DISORDERED BREATHING AMONG NEWBORN INFANTS WITH MYELOMENINGOCELE

Fauziya Hassan[1], Thornton Mason[2], Harlan McCaffery[3], Ronald Chervin[4], Renee Shellhaas[5]
Sleep Disorder Center, Department of Neurology, Division of Pediatric Pulmonology, Department of Pediatrics, University of Michigan [1] The Children's Hospital of Philadelphia [2] Department of Pediatrics, University of Michigan [3] Sleep Disorders Center, Department of Neurology [4] Division of Pediatric Neurology, Department of Pediatrics [5]

Introduction: Myelomeningocele (MMC) is a neural tube defect associated with hindbrain herniation (Chiari II malformation) and respiratory center dysfunction. Prior cross-sectional polysomnographic studies indicated that older children with MMC have an elevated risk of sleep-disordered breathing (SDB), a risk factor for sudden death. Most infants with MMC (78%) had abnormal pneumograms, reported predominantly as central sleep apnea (CSA) and sleep-related hypoventilation (SRH). Pneumograms, however, have significant limitations compared with full polysomnography (PSG).
Methods: The North-American Fetal Therapy Network (NAFTNet) with nine participating sites has collaborated in a prospective study of SDB among infants with MMC. Bedside PSGs were conducted among infants >35 weeks post-menstrual age without supplemental oxygen or respiratory support. PSGs were scored by a pediatric-experienced RPSGT using the American Academy of Sleep Medicine infant sleep staging and pediatric scoring criteria for respiratory events. PSGs were reviewed independently by two board-certified pediatric sleep faculty who then reached diagnostic consensus.
Results: Twenty-eight PSGs were evaluated as an interim analysis from 4 of 9 participating sites. Many (11/28, 39.3%) infants had predominantly frequent hypopneas, which could not be distinguished confidently as central vs. obstructive by two experienced pediatric physicians. The proportions of neonates with CSA (3/28, 10.7%), OSA (6/28, 21.4%) and SRH (1/28, 3.6%) were small by comparison. Only 10/28 infants (35.7%) did not display significant SDB and 14/28 had PSG abnormalities considered clinically concerning. Across all subjects the median [IQR] hypopnea index was 18 [10, 33], central apnea index was 4 [1,7]) and obstructive apnea index was 1.0 [0, 6]. The median [IQR] apnea-hypopnea index was 28 [15, 46].
Conclusion: This ongoing study already provides the largest available cohort of neonates with MMC and PSG data. Predominant hypopneas were far more common than any other classified expression of SDB and were challenging to distinguish as central or obstructive. These data confirm the high frequency of SDB in MMC (64%), suggest that PSG may be an important consideration in neonates with MMC, but highlight that current scoring criteria may not always allow confident separation of central from obstructive SDB processes.
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0532

DEFINING SLEEP ARCHITECTURE IN PEDIATRIC PATIENTS WITH PRADER WILLI SYNDROME

Neha Patel[1], Jenny Lew[1], Gustavo Nino[1], Miriam Weiss[1], Julia Aziz[1]
Children's National Medical Center [1]

Introduction: Prader Willi Syndrome (PWS) is a rare genetic disorder characterized by infantile hypotonia, hyperphagia leading to early childhood obesity, and short stature. PWS patients are at risk for multiple sleep abnormalities including increased risk of obstructive sleep apnea, central sleep apnea, and excessive daytime sleepiness. The limited studies reviewing PSGs in pediatric PWS patients showed varying results. Our study aim is to characterize sleep characteristics in pediatric patients with PWS.
Methods: We conducted a retrospective chart review of pediatric patients with a genetically confirmed diagnosis of Prader Willi Syndrome from 2007 to 2015. In lab polysomnograms were scored using the AASM criteria for pediatric sleep studies. Data was collected on sleep architecture parameters and compared to normative data available for pediatric polysomnography. Given large variability in sleep architecture during infancy, subjects were divided into two age groups (< 1 years old, 1-19 years old).
Results: Fifty-one PSGs were reviewed of which 31 (60.7%) belong to males and 20 (39.3%) belong to females. Forty-one PSGs

were initial studies and 11 were repeat studies. Age at the time of sleep study ranged from 11 days to 19 years old. 8 (15%) subjects were less than 1 years old and 44 (84%) were 1 year and older. For subjects less than one year old, mean sleep onset latency (min) was 21±33.4, REM onset latency (min) 35±27.2, SWS(%) 43.3±10.2, REM(%) 35.3±7.35, arousal index 9.7±11.9, wake after sleep onset (WASO) (min) 59.5±11.4 and sleep efficiency 82.4±9.4. For the 1-19 year age group, mean sleep onset latency (min) was 24.5±23.1, REM onset latency (min) 104± 59.9, SWS (%) 21.4±7.11, REM (%) 21.3±6.9, arousal index 7.3±5.4, WASO (min) 46.5±36.10 and sleep efficiency was 85.7±9.0.

Conclusion: In our population of PWS patients, REM onset latency was not decreased as shown in previous studies. Sleep efficiency was decreased. Other sleep parameters fell within normal range. Additional data on signs of excessive daytime sleepiness and sleep disordered breathing in this population is needed to understand if EDS persists with decreased sleep efficiency and otherwise normal sleep parameters and/or with OSA.

Support (If Any):

0533

AGE-RELATED CHANGES IN SLEEP DISORDERED BREATHING IN PEDIATRIC PRADER WILLI SYNDROME PATIENTS

Neha Patel[1], Nino Gustavo[1], Jenny Lew[1], Miriam Weiss[1], Julia Aziz[1]
Children's National Medical Center [1]

Introduction: Prader Willi Syndrome (PWS) is a rare genetic disorder characterized by infantile hypotonia, hyperphagia leading to early childhood obesity, and multiple sleep abnormalities including increased risk of obstructive sleep apnea, central sleep apnea, and excessive daytime sleepiness. PWS patients have been reported to have a shorter lifespan with respiratory related causes as a common cause of death. Limited data exists on PSGs in pediatric PWS patients. Our aim was to compare respiratory parameters in different age groups.

Methods: We conducted a retrospective chart review of pediatric patients with a genetically confirmed diagnosis of Prader Willi Syndrome from 2007 to 2015. In lab polysomnograms were scored using the AASM criteria for pediatric sleep studies. As central apneas are more commonly seen in infancy with PWS, we categorized patients into < 1 years old and 1-19 years old for comparison.

Results: Fifty-one PSGs were reviewed of which 31 (60.7%) were males and 20 (39.3%) were females. Forty-one PSGs were initial studies and 11 were repeat studies. Age at time of sleep study ranged from 11 days-19 years old. 8 (15%) subjects were < 1 years old and 44 (84%) were 1-19 years old. For patients < 1 year old, mean obstructive hypopnea index (OAHI) was 5.65±3.7, central index (CI)2.2±2.5, mean SpO2(%) 98±.4, SpO2 nadir 86±5.8, and % of time with SpO2 < 90% .16±.314. For patients 1-19 years old, mean (OAHI) was 8.7±13.4, (CI) 0.8±1.25, mean SpO2 96±3.0, SpO2 nadir 81±.10, and % of time with SpO2 < 90% .03±.10. The OAHI trended up in the 1-19 year old group but results were not significant, CI decreased in the 1-19 year old group 2.2 vs 0.8 p < .04.

Conclusion: Our study demonstrated similar results to prior studies in pediatric PWS with an increase in OAHI and decrease in CI with age. SPO2 nadir decreased in the 1-19 year old group but these results were not significant. Ongoing research at our center is looking at possible contributing factors including BMI percentile, GH therapy, and adenotonsillectomy to better understand our findings.

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0534

DYSPHAGIA SEVERITY IS ASSOCIATED WITH WORSE SLEEP DISORDERED BREATHING IN INFANTS WITH TRISOMY 21

Yeilim Cho[1], Lourdes DelRosso[2], Michelle Sobremonte-King[3]
University of Washington Division of Pulmonary, Critical Care and Sleep Medicine [1] University of Washington, Seattle Children's Hospital, Division of Pulmonary and Sleep Medicine [2] University of Washington School of Medicine, Seattle Children's Hospital Division of Pulmonary and Sleep Medicine [3]

Introduction: Hypotonia is common in infants with Trisomy 21. This can cause masticatory and oropharyngeal muscle weakness increasing the risk for dysphagia and sleep disordered breathing. Data describing the occurrence of dysphagia and sleep disordered breathing in infants with Trisomy 21 is limited. This study aims to determine the frequency and severity of dysphagia and its relationship to polysomnogram parameters in infants with Trisomy 21.

Methods: Retrospective chart review of patients with Trisomy 21 <12 months old that underwent polysomnography at Seattle Children's Hospital between October 1, 2015-August 23, 2021. Data collected included: sex, age, presence of dysphagia, recommended thickener type and polysomnographic data.

Results: A total of 526 polysomnograms in patients with Trisomy 21 were performed. Forty-one studies were identified in <12 months old. Results in mean ± SD showed: age 6.5 months + 3, 66% were male and 73% were diagnosed with dysphagia through a video fluoroscopic swallow study. In those with dysphagia, 16% can tolerate thin liquids, 20% prescribed nectar-thick, 7% prescribed honey-thick and 57% were G-tube dependent. In patients with dysphagia compared to those without dysphagia: there was higher total AHI of 43.3 +/- 35.3 vs. 22.6 +/- 10.6 (p=0.006), oAHI of 39.7 +/- 35.5 vs. 17.2 +/- 11.6 (p=0.004), CAI of 3.4 +/- 3.4 vs. 3.4 +/- 1.8 (p=0.11), oxygen saturation nadir of 78.6 +/- 10.6 vs. 83.1 +/- 6.6 (p=0.11) and percentage total sleep time TcCO2 >50 mmHg of 44.6 +/- 42.6 vs. 31 +/- 40.3 (p=0.44). Worse dysphagia was positively correlated with a higher oAHI (r=0.38, p=0.03).

Conclusion: There is a high incidence of dysphagia and sleep disordered breathing in infants with Trisomy 21. Dysphagia severity correlated with oAHI severity. Dysphagia in OSA can be due to the sensory and motor changes of the pharynx with impaired swallow-breathing mechanism. Chronic microaspiration can also result in decreased pulmonary reserve from lower airway inflammation or lung parenchymal disease, which may lead to worse sleep disordered breathing. Current guidelines suggest screening at school age or when there are clinical symptoms of OSA in Trisomy 21. However, results suggest the need to evaluate and intervene earlier especially in infants with dysphagia.

Support (If Any):

0535

PREVALENCE OF VARIOUS FORMS OF SLEEP DISORDERED BREATHING IN INFANTS WITH DOWN SYNDROME.

Yeilim Cho[1], Younghoon Kwon[2], Chris Ruth[1], Samuel Cheng[3], Lourdes DelRosso[1]
University of Washington School of Medicine, Seattle Children's Hospital Division of Pulmonary and Sleep Medicine [1] University of Washington [2] Seattle Children's Hospital, Seattle WA [3]

Introduction: Children with Down Syndrome (DS) are at high risk of sleep disordered breathing (SDB). Undiagnosed SDB in