The National Healthy Sleep Awareness Project involves a partnership between the American Academy of Sleep Medicine, Center for Disease Control and Sleep Research Society. The long term goal of the project is to promote improved sleep health in the United States. The project will increase public awareness of the importance of healthy sleep. It also will promote the treatment and prevention of sleep disorders.

Pediatric Populations at High Risk for Sleep Apnea

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The prevalence of sleep apnea in children is estimated to be 1.2% to 5.7%,[1] but in certain groups, is much higher. Recognition of sleep apnea is important in children, as it is associated with behavioral, neurocognitive, learning and cardiovascular problems.[2-4] Importantly, these adverse effects are reversible with treatment of sleep apnea such as adenotonsillectomy or positive airway pressure therapy.[5-7] Sleep apnea can be considered as either obstructive or central in nature, and different underlying factors may predispose some children toward each type.

Obstructive sleep apnea (OSA) is due to upper airway collapse during sleep despite persistent respiratory effort. Sleep apnea in children is much more commonly obstructive than central. In otherwise healthy pre-school age children, this obstruction is frequently related to adenotonsillar hypertrophy, and responds well to treatment with adenotonsillectomy.[8] In addition to possible adenotonsillar hypertrophy, children with other underlying disorders may be at increased risk for OSA through mechanisms including airway narrowing, other causes of airway obstruction and decreased airway tone. Examples of disorders associated with airway narrowing include Apert syndrome or Treacher-Collins syndrome. Macroglossia, seen in conditions such as Beckwith-Wiedemann, or glossoptosis, seen in Pierre Robin sequence, may also contribute to obstruction. Children with Down syndrome are well known to be at increased risk for OSA,[9] and this is likely due to a combination of low airway tone, airway narrowing (due to mid-face hypoplasia) and obstruction related to relative macroglossia. An additional group at high risk is children with mucopolysaccharidoses, due to deposition of glycosaminoglycans obstructing the airway.[10] In general, there should be a high suspicion of obstructive sleep apnea in all children with craniofacial abnormalities, particularly those associated with mid-face hypoplasia or retrognathia/micrognathia. Treatment of OSA in these populations must be tailored to the

underlying disorder. Standard treatments such as adenotonsillectomy or positive airway pressure therapy may be effective in some children, however some disorders may respond well to surgical and orthodontic procedures that lead to increased airway size. Examples of this include rapid mandibular expansion, palate expansion or mandibular distraction osteogenesis. In certain cases, tracheostomy may be necessary to treat the OSA.

Central sleep apnea (CSA) occurs when there is an absence of respiratory effort, but no airway obstruction. This may be due to a central nervous system problem, or due to muscular weakness affecting the diaphragm and other respiratory muscles. Central nervous system control of respiration is mediated through the dorsal respiratory group (located in the medulla) and ventral respiratory group (extending from the medulla to the first cervical segment of spinal cord). Any injury in this area can therefore disrupt respiratory control and result in CSA. Arnold-Chiari malformations are a relatively common cause of this, due to impingement of the medulla. Children found to have type 1 Arnold-Chiari malformations may present with headaches, but are also at risk for central sleep apnea, and should be screened for related symptoms such as morning headaches, un-refreshing sleep or daytime hypersomnolence. Children with myelomeningocele are at risk for CSA related to type 2 Arnold-Chiari malformation.[11] Children with myelomeningocele also appear to be at an increased risk for OSA as well, thought to be related to impaired central nervous system control of airway musculature. Children with achondroplasia have a similar high risk of CSA due to brainstem compression from foramen magnum bony stenosis, and are at high risk for OSA as well due to mid-face hypoplasia.[12] Children with muscular dystrophy syndromes are at high risk for CSA, OSA and sleep-related hypoventilation. This is due to underlying muscle weakness that affects both respiratory muscles, as well as airway musculature. The onset of apnea is dependent upon the underlying dystrophy, with some children presenting with apnea in infancy (i.e. spinal muscular atrophy[13]), and others may not develop sleep apnea until teenage years(Duchenne muscular dystrophy[14]). Treatment for children with CSA is generally PAP therapy, however children with brainstem compression (such as an Arnold-Chiari malformation) may benefit from neurosurgical procedures.

In general, there should be a high suspicion of sleep apnea in all children with craniofacial malformations, brainstem abnormalities, or muscular weakness. Given the known benefits of treatment of sleep apnea[5-7] in children, early recognition of

sleep apnea in these at-risk children is likely to result in significant improvements in their long-term health.

REFERENCES

1. Marcus CL, Brooks LJ, Draper KA, et al. Diagnosis and management of childhood obstructive sleep apnea syndrome. *Pediatrics*. 2012;130(3):e714–e755.
2. Kaemingk KL, Pasvogel AE, Goodwin JL, et al. Learning in children and sleep disordered breathing: findings of the Tucson Children's Assessment of Sleep Apnea (tuCASA) prospective cohort study. *J Int Neuropsychol Soc*. 2003;9(7):1016–1026.
3. Gozal D. Sleep-disordered breathing and school performance in children. *Pediatrics*. 1998;102(3 Pt 1):616–620.
4. Amin RS, Kimball TR, Kalra M, et al. Left ventricular function in children with sleep-disordered breathing. *Am J Cardiol*. 2005;95(6):801–804.
5. Friedman BC, Hendeles-Amitai A, Kozminsky E, et al. Adenotonsillectomy improves neurocognitive function in children with obstructive sleep apnea syndrome. *Sleep*. 2003;26(8):999–1005.
6. Montgomery-Downs HE, Crabtree VM, Gozal D. Cognition, sleep and respiration in at-risk children treated for obstructive sleep apnoea. *Eur Respir J*. 2005;25(2):336–342.
7. Yuan HC, Sohn EY, Abouezzeddine T, et al. Neurocognitive functioning in children with obstructive sleep apnea syndrome: a pilot study of positive airway pressure therapy. *J Pediatr Nurs*. 2012;27(6):607–613.
8. Brietzke SE, Gallagher D. The effectiveness of tonsillectomy and adenoidectomy in the treatment of pediatric obstructive sleep apnea/hypopnea syndrome: a meta-analysis. *Otolaryngol Head Neck Surg*. 2006;134(6):979–984.
9. de Miguel-Diez J, Villa-Asensi JR, Alvarez-Sala JL. Prevalence of sleep-disordered breathing in children with Down syndrome: polygraphic findings in 108 children. *Sleep*. 2003;26(8):1006–1009.
10. Moreira GA, Kyosen SO, Patti CL, Martins AM, Tufik S. Prevalence of obstructive sleep apnea in patients with mucopolysaccharidosis types I, II, and VI in a reference center. *Sleep Breath*. 2014;18(4):791–797.
11. Kirk VG, Morielli A, Gozal D, et al. Treatment of sleep-disordered breathing in children with myelomeningocele. *Pediatr Pulmonol*. 2000;30(6):445–452.
12. Julliand S, Boule M, Baujat G, et al. Lung function, diagnosis, and treatment of sleep-disordered breathing in children with achondroplasia. *Am J Med Genet A*. 2012;158A(8):1987–1993.
13. Testa MB, Pavone M, Bertini E, Petrone A, Pagani M, Cutrera R. Sleep-disordered breathing in spinal muscular atrophy types 1 and 2. *Am J Phys Med Rehabil*. 2005;84(9):666–670.
14. Sawnani H, Thampratankul L, Szczesniak RD, Fenchel MC, Simakajornboon N. Sleep disordered breathing in young boys with Duchenne muscular dystrophy. *J Pediatr*. 2015;166(3):640–645. e1.

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