Adenotonsillectomy may help resolve obstructive sleep apnea in children with Prader-Willi syndrome

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(Medical Xpress)—Children with Prader-Willi syndrome may receive relief from sleep disorders after undergoing an adenotonsillectomy, suggests a new study from Nationwide Children's Hospital published in the November print issue of the *Archives of Otolaryngology-Head and Neck Surgery*.

"**Patients** with Prader-Willi syndrome are at risk for **sleep disordered breathing** as growth hormone commonly used to treat their condition can cause the tonsils and adenoids to enlarge," said the study's lead author Kris Jatana, MD, FAAP, with Otolaryngology Head & Neck Surgery at Nationwide Children's.

"**Growth hormone** in a low dose is extremely beneficial to most patients with Prader-Willi syndrome," said David Repaske, PhD, MD co-director of the **Prader Willi Center** and chief of Endocrinology, Metabolism & Diabetes at Nationwide Children's. "This beneficial effect has nothing to do with growth, but rather, it markedly improves patients' ability to eat, sit, stand, walk or run depending on their developmental stage and due to a positive effect on their low muscle tone."

At Nationwide Children's, Prader-Willi patients undergo an annual **sleep study** and are evaluated for potential **adenotonsillectomy** if obstructive apnea events are present. To evaluate the efficacy of adenotonsillectomy
in the treatment of sleep apnea in Prader-Willi syndrome, investigators at Nationwide Children's performed a retrospective chart review. Thirteen patients met the study criteria and were categorized based on severe, moderate or mild apnea/hypopnea indexes and obstructive hypoxia.

Findings showed that 89 percent of the patients with mild-to-moderate obstructive sleep apnea or obstructive hypoventilation normalized after receiving adenotonsillectomy. Of the four children with severe obstructive sleep apnea, two normalized after surgery and two continued to have severe apneas.

"These findings suggest that adenotonsillectomy is effective in most children with Prader-Willi syndrome who demonstrate mild to moderate obstructive sleep apnea, but may not be curative in children with severe forms of the condition," said Dr. Jatana, also a faculty member at The Ohio State University College of Medicine.

Dr. Jatana and his team of investigators stress that patients should receive a repeat sleep study six-to-eight weeks postoperatively since an increase in central apneas can occur in some Prader-Willi children after upper airway surgical intervention.

Prader-Willi Syndrome is treated at Nationwide Children's by a multidisciplinary team which emphasizes early intervention and a proactive approach to assessment of and treatment for the possible complications that can be associated with PWS. The team includes an endocrine nurse coordinator, endocrinologists, a geneticist, a genetic counselor and a dietitian that attend each PWS Clinic. The team collaborates with clinicians from a number of supporting disciplines, and patients are referred to Ophthalmology, ENT, Sleep Medicine, Behavioral Health, Urology, Physical/Occupational/Speech Therapy and Dermatology as needed.