International Journal of Pediatric Otorhinolaryngology

# Adenotonsillectomy success for treating obstructive sleep apnea in children with Prader-Willi syndrome

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Volume 192, May 2025, 112305

https://doi.org/10.1016/j.ijporl.2025.112305Get rights and content

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## Highlights

- Children with Prader-Willi Syndrome have high risk of obstructive sleep apnea.
- Sleep apnea in these patients is often multifactorial.
- Adenotonsillectomy has lower success rates for sleep apnea in these patients.
- A multidisciplinary team should be used to properly address OSA in PWS patients.

## Abstract

### Background

Prader-Willi syndrome (PWS) is a rare genetic disorder that can increase risk of pediatric obstructive sleep apnea (OSA), caused by the combination of increased viscosity of secretions, craniofacial abnormalities, hypotonia, and obesity. While first-line treatment of pediatric OSA is typically adenotonsillectomy, the complex pathophysiology of OSA in PWS patients may lead to less success with this therapy.

### Methods

The TriNetX database was queried for patients 18 years old or younger based on the diagnoses of PWS and OSA and the surgical interventions of adenotonsillectomy, tonsillectomy, and adenoidectomy. The primary endpoint was the removal of the diagnosis of OSA 6 months postoperatively. Pediatric patients without PWS were used as a control. Secondary endpoints were the risk of OSA with common medical interventions for children with PWS.

### Results

A total of 2163 patients were found to have PWS, with 1035 (47 %) diagnosed with OSA. PWS patients undergoing surgery had a total success rate of 39.0 %, compared to 79.6 % in controls ($p < 0.001$). Total success for these surgeries was also significantly lower compared to controls matched by demographics and obesity status (36.8 % versus 82.1 %, $p < 0.001$). Use of growth hormone (RR 1.43, $p < 0.001$) and testosterone (RR 1.39, $p < 0.001$) were both associated with increased risk of OSA.

### Conclusions

Adenotonsillectomy has significantly lower rates of success at treating pediatric OSA in patients with PWS. These patients would likely benefit from multidisciplinary care to treat their OSA and mitigate the effects of untreated disease, and further studies determining best practices for caring for these patients are necessary.

## Introduction

Prader-Willi Syndrome (PWS) is a multisystem genetic disorder with an estimated prevalence of 1–3 per 100,000 [1]. Common symptoms include severe hypotonia, feeding difficulties in infancy, motor and language delays, behavioral disorders, hypogonadism, and growth hormone (GH) deficiency, with a hallmark symptom of excessive eating and the development of morbid obesity. These patients commonly develop sleep disorders such as reduced rapid eye movement (REM) latency, altered sleep architecture, oxygen desaturations, and central and obstructive sleep apnea [[2], [3], [4]]. It is estimated that approximately 80 % of patients with PWS have obstructive sleep apnea (OSA) [3].

The development of OSA in children with PWS is often multifactorial, with influence from obesity, hypotonia, micrognathia, small oropharynx, and viscous secretions [3]. Considering the high prevalence of hormonal disorders such as GH deficiency, hypogonadism, hypoadrenalism, and hypothyroidism in these patients, recent evidence has driven theories that PWS pathologies are related to hypothalamic dysfunction [4]. As the hypothalamus is important for respiratory control and sleep/wake regulation, PWS patients often have abnormal ventilatory responses to hypoxia and hypercapnea [2,3]. This leads to central sleep apnea (CSA) in infancy; while this typically improves with time, they continue to have difficulty with respiratory regulation. With all of these difficulties, treatment of OSA in children with PWS is often more complicated than in otherwise healthy children.

As untreated sleep apnea can lead to major cardiovascular and respiratory complications, it is important to understand the disease process and how to effectively treat the condition [[5], [6], [7]]. In healthy children, adenotonsillectomy (T&A) is considered the first-line and most effective treatment modality for OSA [8]. However, the complex nature of sleep apnea in PWS patients has led to lower reported success rates with this treatment. As PWS is an uncommon condition, few studies reported on this outcome, with the largest being a meta-analysis of 41 patients [8]. Here, we report on a study utilizing a large, federated database examining the success of T&A in children with PWS.

## Methods

The TriNetX research network is a federated multicenter research network that provides real-time access to a deidentified data set from the electronic health records of participating healthcare organizations. The database incorporates all diagnoses and treatments from a patient's medical records regardless of the provider inputting them, as long as the provider is utilizing a system from which the database pulls information. Diagnoses can be searched in the database through International

## Results

A total of 2163 patients with PWS were found, with 1035 (47 %) having a diagnosis of OSA. This is compared to 404,758 out of 34,200,721 (1 %) non-PWS children having a diagnosis of OSA. Of the 840 PWS patients with an included BMI ICD-10 code, 643 (77 %) were classified as overweight or obese. Of patients diagnosed with OSA, patients with PWS were diagnosed with OSA at a younger age compared to those without PWS (5.4 vs. 7.3, $p < 0.001$). General characteristics of PWS and non-PWS patients are

## Discussion

Children with Prader-Willi Syndrome present a unique challenge to healthcare providers treating sleep apnea. Due to the multifactorial etiology of OSA in these patients, T&A may not be as successful at treating the condition compared to OSA in otherwise healthy children. Considering the cardiovascular and respiratory implications of untreated OSA, it is important to understand trends in risk factors and treatments for OSA in this population. However, due to the rarity of PWS, there is a paucity

Conclusions

Adenotonsillectomy has significantly lower rates of success at treating pediatric OSA in patients with PWS. Other common interventions for children with PWS such as GH therapy may overall improve the health of these patients but have mixed effects on OSA. These patients would likely benefit from multidisciplinary care to treat their OSA and mitigate the effects of untreated disease, and further studies determining best practices for caring for these patients are necessary.

Funding

This study received no funding

Declaration of competing interest

No authors have any relevant financial interests to disclosure.

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