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Efficacy and Safety of Adenotonsillectomy for Pediatric Obstructive Sleep Apnea in Prader-Willi Syndrome

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Objectives: We performed a retrospective analysis in a tertiary care children's hospital to evaluate the efficacy and safety of adenotonsillectomy for the treatment of pediatric obstructive sleep apnea (OSA) in Prader-Willi syndrome (PWS).

Methods: We studied all PWS patients who underwent adenotonsillectomy to treat OSA from January 1, 2004, to December 31, 2005. The main outcome measures were 1) preoperative and postoperative full overnight polysomnography and 2) postoperative complications.

Results: Three PWS patients were identified, including female twins and 1 boy. All patients had preoperative evidence of OSA without central apnea. Resolution of OSA after adenotonsillectomy was variable. The patient with the highest body mass index and tonsil size had the least residual OSA after adenotonsillectomy. No perioperative complications or adverse events were observed.

Conclusions: Adenotonsillectomy did not consistently improve OSA in this population of patients with PWS. No perioperative complications were noted. Postoperative polysomnography should be considered for evaluation of possible residual OSA, as additional interventions may be warranted.

Key Words: adenotonsillectomy, apnea, obstructive sleep apnea, pediatric sleep apnea, Prader-Willi syndrome.

INTRODUCTION

Prader-Willi syndrome (PWS) is an uncommon genetic disorder that afflicts an estimated 1 in 10,000 to 1 in 25,000 live births.[1,2] The majority of patients have a deletion in the paternally derived chromosome 15.[3] The clinical manifestations of the disorder are numerous and vary with age, often making diagnosis difficult.[1] Failure to thrive and hypotonia are features characteristic of the neonatal period. By infancy and early childhood, patients may exhibit developmental delay, hypogonadism, and hyperphagia with resulting obesity. Behavioral dysfunction becomes apparent by adolescence.[4]

Sleep-disordered breathing is a reported finding in children with PWS (Table 1). It often occurs by early childhood and is accompanied by daytime hypersomnolence, snoring, and frequent nocturnal arousals.[5] Sleep-disordered breathing may manifest as any combination of obstructive sleep apnea (OSA), alveolar hypoventilation, or restrictive lung disease. Risk factors for the development of sleep-disordered breathing in these patients include hypotonia, obesity, and, possibly, adenotonsillar hypertrophy.[6,7] Although adenotonsillectomy is widely performed in the management of obstructive sleep apnea in otherwise normal pediatric patients, its use in PWS has been inadequately investigated.[8] Given the paucity of studies investigating the efficacy and safety of adenotonsillectomy in this patient group, the authors aimed to evaluate these parameters in this select patient group.

PATIENTS

A retrospective chart review was performed on all patients with PWS who underwent adenotonsil-

TABLE 1. CHARACTERISTICS OF SLEEP-DISORDERED BREATHING IN PRADER-WILLI SYNDROME

Daytime hypersomnolence
Restless sleep
Hyperactivity
Witnessed apneas
Snoring
Reduced rapid eye movement latency
Obstructive sleep apnea
Alveolar hypoventilation
Restrictive lung disease

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TABLE 2. PATIENT CHARACTERISTICS AND RESULTS

	Patient 1		*Patient 2*		*Patient 3*	
	Preop	*Postop*	*Preop*	*Postop*	*Preop*	*Postop*
Age (y)	3		3		12	
Apnea-Hypopnea Index	13.1	13.3	6.3	2.4	49.8	1.57
Central apneas (per hour)	0.7	0	0	0.8	0	0
Oxygen saturation (%)						
Range	82.5-100	71-100	94.4-100	77.5-100	54.7-100	91.1-100
Mean	98.6	97.7	98.9	98.2	96.5	98
Tonsil grade	2+		2+		4+	
Height (m)	0.9		0.88		1.45	
Weight (kg)	14.8		13.9		87.3	
Body mass index	18.27		17.95		41.52	

lectomy between January 1, 2004, and December 31, 2005. Outcome parameters included a comparison of preoperative and postoperative full overnight polysomnography (PSG) reports and the occurrence of any postoperative complications. All postoperative PSG was performed at least 6 months after surgical intervention.

Three PWS patients, including female twins (patients 1 and 2) and 1 boy (patient 3), were identified. None of the patients were receiving growth hormone. The body mass indices for patients 1 and 2 were 18, and that of patient 3 was 42. The preoperative PSG results for patients 1, 2, and 3 included an obstructive Apnea-Hypopnea Index of 13.1, 6.3, and 49.8 events per hour; a percentage of recorded time with oxygen saturation of 90% or less of 0.2%, 0%, and 13.6%; and a nadir oxygen saturation of 83%, 94%, and 55%, respectively. The postoperative PSG results for patients 1, 2, and 3 included an obstructive Apnea-Hypopnea Index of 13.3, 2.4, and 1.6 events per hour; a percentage of recorded time with oxygen saturation of 90% or less of 4.4%, 0.1%, and 0%; and a nadir oxygen saturation of 71%, 78%, and 91%, respectively. Both before and after operation, the mean oxygen saturation was greater than 97% and no central apneic events were noted. No postoperative complications or adverse events were observed. The patients required less than 24 hours of postoperative supplemental oxygen. By postoperative day 3, all patients were discharged. Table 2 summarizes the patient results.

DISCUSSION

The disease characteristics predispose patients with PWS to OSA. Facial dysmorphisms, including micrognathia, alter the pharynx.[9] Morbid obesity, hypotonia, and increased secretions are other risk factors.[6,7,9] The true prevalence of OSA in PWS is unknown.[6] Various investigators have attempted to define this. However, small numbers of subjects, variable inclusion criteria, and inconsistent methods of PSG have precluded an accurate estimate.

Early diagnosis of sleep-disordered breathing, in particular OSA, in PWS patients may help prevent complications such as cor pulmonale.[10] Given the ill-defined prevalence of OSA in PWS and the difficulty in predicting OSA with clinical history alone, the authors advocate that all patients with PWS receive PSG testing, especially if surgical interventions aimed at treating OSA are being considered.

The sleep-disordered breathing in PWS is multifactorial in cause. Unlike in otherwise normal children, adenotonsillar hypertrophy may only be a small contributing factor in the development of OSA in PWS. Other factors, such as obesity, hypotonia, and altered anatomy, may have a greater impact on sleep-disordered breathing. As such, the use of adenotonsillectomy in patients with PWS must be considered carefully, weighing the safety of the surgery in this patient population against its efficacy. Although no patient in this study experienced perioperative complications, adenotonsillectomy in children with PWS is not without risk. Additional-

TABLE 3. COMPREHENSIVE EVALUATION OF PATIENTS WITH PRADER-WILLI SYNDROME BEING CONSIDERED FOR ADENOTONSILLECTOMY

History
Daytime hypersomnolence, snoring, restless sleep, witnessed apnea
Sleep pattern
Physical examination
Body mass index
Adenotonsillar size
Assessment of anatomy and hypotonia with flexible laryngoscopy
Cardiovascular evaluation
Consideration of cor pulmonale
Pulmonary evaluation
Consideration of restrictive lung disease or alveolar hypoventilation
Polysomnography

ly, it has been demonstrated that patients with obesity or significant cardiopulmonary histories are at increased risk during this surgery.[11] Therefore, the authors suggest that the consideration of performing adenotonsillectomy in PWS is one that should not be taken lightly.

Adenotonsillectomy in PWS requires a comprehensive evaluation of the patient (Table 3). With the other contributing factors to OSA, it is unlikely that there will be complete resolution. As such, the authors advocate a postoperative overnight observation and postoperative PSG. In our series, patient 1 had no improvement in PSG results after adenotonsillectomy. This finding highlights the authors' point that OSA in PWS is multifactorial, beyond just adenotonsillar hypertrophy. Similarly, in a previously published series of 5 pediatric patients, not all PWS patients had resolution of OSA with adenotonsillectomy.[8]

The management options aside from adenotonsillectomy include weight loss, which our patients had limited success with, nasal continuous positive airway pressure, and tracheostomy.[12,13] These treatment options can be considered if adenotonsillectomy is not performed or if OSA symptoms persist after adenotonsillectomy. The patients in this study with no resolution of OSA were referred for nasal continuous positive airway pressure.

CONCLUSIONS

Obstructive sleep apnea is a reported clinical manifestation of PWS and is often multifactorial in cause. Before considering adenotonsillectomy, the authors urge surgeons to consider a preoperative PSG and the possible increased risk of perioperative complications. If adenotonsillectomy is performed, postoperative PSG may be considered for evaluation of possible residual OSA, as additional interventions may be warranted. Given the paucity of literature regarding OSA in PWS, larger-scale studies are needed.

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