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Surgical outcomes and sleep endoscopy for children with sleep-disordered breathing and hypotonia

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ABSTRACT

Objective: To study the efficacy of surgical management for obstructive sleep apnea (OSA) syndrome in children with hypotonia, and to identify common anatomic sites of airway obstruction.
Methods: Retrospective chart review of polysomnographic parameters and quality of life instrument scores for seventy eight children with hypotonia who underwent surgical intervention for sleep-disordered breathing at two tertiary children's hospitals, and analysis of drug-induced sleep endoscopy data using a previously validated scoring system.
Results: Children undergoing surgical intervention had baseline severe OSA with a statistically significant improvement in apnea-hypopnea index from 23.6 to 11.1 after surgery, but persistent severe OSA. OSA-18 sleep-related quality of life measurement and overall quality of life score showed statistically and clinically significant improvements, from 72.0 to 43.4 and from 5.3 to 7.6 respectively. Sleep endoscopy showed an average obstructive score of 7.2/15 (n = 39), with multi-level obstruction in 49% of children. Greater than 50% obstruction was observed at the tongue base in 64% of patients, velum in 46%, lateral pharyngeal wall in 38%, supraglottis in 38%, and adenoid in 23%.
Conclusion: OSA syndrome is challenging to treat in hypotonic children. Severe residual OSA is common after surgical intervention, but improvement in quality of life is clinically and statistically significant. The tongue base is the most common site of persistent airway obstruction. Drug-induced sleep endoscopy can identify sites of airway obstruction and may aid in surgical planning for high-risk patients.

1. Introduction

Childhood obstructive sleep apnea syndrome (OSAS) is characterized by partial or complete airway obstruction during sleep resulting in repetitive arousals and/or oxygen desaturations [1]. In most cases, this common condition, estimated to affect 1–4% of children, is effectively treated surgically in order to avert the neurocognitive, behavioral, cardiovascular, and inflammatory sequelae of untreated OSAS [1–6]. However, patients with certain risk factors are at high risk of disease refractory to surgery. In particular, a comorbid diagnosis such as asthma, obesity, or Down syndrome greatly increases the likelihood of severe refractory OSAS [5,7–11]. Hypotonia is considered a risk factor for relative surgical failure, but this has not been definitively shown [11], with a paucity of data in the literature describing the underlying pathophysiology and the efficacy of surgical management of OSAS in hypotonic children.

We present a retrospective cross-sectional analysis of children with hypotonia who were surgically managed for sleep-disordered breathing at two tertiary children's hospitals. We sought to describe treatment course and residual disease burden through evaluation of OSA symptoms and sleep-related quality of life, and to gain a better understanding of the pathophysiology and severity of airway obstruction at multiple anatomic levels utilizing a previously validated sleep endoscopy scoring system [12].

2. Methods

2.1. Patient selection and data collection

This study was approved by the institutional review boards of the University of California, San Francisco and Seattle Children's

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[1] Drs. Park and Chan had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis.

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Hospital. The investigators adhered to policies for protection of human subjects as prescribed in 45 CFR 46. Waiver of written informed consent was obtained and approved by both institutional review boards. Children with a clinical diagnosis of hypotonia who underwent surgical procedures for sleep-disordered breathing by attending pediatric otolaryngologists at the UCSF Benioff Children's Hospital from 2002 through 2014 and from Seattle Children's Hospital from 2011 to 2013 were collected in quality-improvement databases. Databases were queried retrospectively at the time of the study.

Primary inclusion criteria were documentation of a surgical procedure for treatment of sleep-disordered breathing and a diagnosis of hypotonia. During chart review, particular attention was taken to multidisciplinary evaluation by pediatricians, pediatric neurologists, and medical geneticists supporting a diagnosis of hypotonia on physical exam, thus selecting for children with syndromic and non-syndromic hypotonia and neuromuscular dysfunction. Patients were excluded if evaluation for sleep-disordered breathing had been conducted (including polysomnography, sleep endoscopy, or direct laryngoscopy and bronchoscopy), but no treatment procedure had been performed. No additional exclusion criteria were applied, in order to improve study generalizability.

During chart review for 78 included patients, demographic data, operative details, polysomnographic indices, drug-induced sleep endoscopy findings, and OSA-18 quality of life questionnaire data were collected.

2.2. OSA-18 questionnaire for sleep-related quality of life

The OSA-18 questionnaire is an 18-item seven point Likert scale survey measuring quality of life in five domains: sleep disturbance, physical suffering, emotional symptoms, daytime function, and caregiver concern [13]. The OSA-18 index, or summary score, is the sum of all item scores, with a minimum of 18 and a maximum of 126 points and a value of 60 or above considered abnormal. A visual analog scale item asking caregivers to rate the child's overall quality of life (scored 0–10 with 10 representing the best possible quality of life) was also on the questionnaire. The questionnaire was administered at clinic visits and scores recorded in the electronic medical record.

2.3. Pediatric polysomnography

Polysomnograms were performed at the sleep laboratory at Seattle Children's Hospital or at the UCSF Benioff Children's Hospital Pediatric Sleep Laboratory. These were scored according to standard American Academy of Sleep Medicine scoring criteria for pediatric polysomnography [14]. The apnea-hypopnea index (AHI) was defined as the total number of apneas and hypopneas averaged per hour of total sleep time. The oxygen saturation nadir was defined as the lowest oxygen saturation recorded during an obstructive event. AHI and oxygen saturation nadir values were obtained from the primary reports of polysomnograms. Obstructive sleep apnea syndrome was categorized by AHI as absent (<1), mild (1–5), moderate (5–10), or severe (>10).

2.4. Validated scoring system for drug-induced sleep endoscopy

Transnasal flexible fiber-optic drug-induced sleep endoscopy (DISE) was performed in the operating room with patients positioned supine and spontaneously ventilating under anesthesia. The standard regimen for DISE at both institutions is total IV anesthesia via propofol drip. As previously described, videos were evaluated for airway obstruction on a 0–3 scale at five anatomic sites: adenoids, velum, lateral pharyngeal walls, tongue base, and supraglottis [12]. Higher total obstructive scores and multilevel obstruction – defined as significant obstruction (50–99%, score ≥ 2) within both the upper airway complex (adenoid, velum, or lateral pharyngeal wall) and the lower airway complex (tongue base or supraglottis) – are correlated with worsened polysomnographic parameters [12].

2.5. Statistical analysis

Comparisons of paired nonparametric data (including AHI and oxygen saturation nadir) were performed with the Wilcoxon rank-sum test. Unpaired nonparametric data were compared with the Mann-Whitney rank-sum test. Parametric data were compared with the *t*-test. Binomial data were compared with the Fisher exact test. Statistical significance was set at $p < 0.05$.

3. Results

3.1. Patient population

78 patients with hypotonia underwent surgical procedures for sleep-disordered breathing. Study population characteristics are given in Table 1. The most common comorbidities were Down syndrome (n = 24, 31%), developmental delay (n = 22, 28%), and cerebral palsy (n = 12, 15%). Other comorbid diagnoses included Trisomy 13, partial Trisomy 18, chromosome 1q43q44 duplication, unspecified chromosomal abnormality, generalized muscle weakness, laryngomalacia, epilepsy, agenesis of the corpus callosum,

Table 1
Demographic and clinical characteristics of the study patients.[a]

Characteristic		
Total patients, No.	78	
Age, y	5.3	(0.46; 4.4–6.2)
Pre-op AHI (N = 48)	20.1	(3.4; 13.2–27.0)
Pre-op O_2, % (N = 48)	80.5	(1.7; 77.0–84.0)
Post-op AHI (N = 38)	11.8	(2.8; 6.2–17.4)
Post-op O_2, % (N = 38)	83.9	(1.2; 81.5–86.3)
Pre-op OSA-18 (N = 23)	72.0	(5.2; 61.9–82.9)
Pre-op OSA-18 overall QOL (N = 23)	5.3	(0.42; 4.4–6.2)
Post-op OSA-18 (N = 14)	43.9	(6.0; 31.0–56.8)
Post-op OSA-18 overall QOL (N = 14)	7.6	(0.59; 6.3–8.9)
Mean# procedures per patient	1.7	(0.15; 1.4–2.0)
Patients with 1 procedure, No. (%)	56	(72)
Patients with 2 procedure, No. (%)	8	(10)
Patients with >2 procedures, No. (%)	14	(18)
Procedures received		
Adenotonsillectomy, No. (% of patients)	62	(80)
Supraglottoplasty, No. (% of patients)	14	(18)
Revision adenoidectomy, No. (% of patients)	10	(13)
Turbinate reduction, No. (% of patients)	10	(13)
Lingual tonsillectomy, No. (% of patients)	9	(12)
Adenoidectomy, No. (% of patients)	6	(8)
Tonsillectomy, No. (% of patients)	3	(4)
Revision tonsillectomy, No. (% of patients)	3	(4)
Genioglossus advancement, No. (% of patients)	3	(4)
Inter-arytenoid injection, No. (% of patients)	1	(1)
Palatoplasty, No. (% of patients)	1	(1)
Common comorbidities		
Down syndrome, No. (%)	24	(31)
Developmental delay, No. (%)	21	(27)
Cerebral palsy, No. (%)	12	(15)

Abbreviations: O_2, oxygen saturation; post-op, postoperative; pre-op, preoperative.
[a] Unless otherwise indicated, data are reported as mean (SEM; 95% CI) values.