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Airway obstruction in children with cerebral palsy: Need for tracheostomy?

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ABSTRACT

Objective: To examine the progress of the airway obstruction over time in children with cerebral palsy (CP) and the timing of any interventions.
Methods: The medical notes of patients with CP younger than 16 years admitted with airway obstruction to a tertiary referral Pediatric Otolaryngology Center from 2006 to 2012 were retrospectively reviewed. The gender, age of referral, co-morbidities, type of surgical intervention and age this was performed and the time interval between sequential surgeries were documented.
Results: Fifteen children with CP and airway obstruction were admitted, eight boys and seven girls with an average age of referral 8 years (range 3–13.3 years). Adenotonsillectomy was performed in 11/15 patients at a mean age of 9.1 years (range 4.5–14 years). Tracheostomy was performed in 8/15 children at an average age of 11.6 years (range 7.5–15 years). Seven out of 11 patients having undergone adenotonsillectomy, required tracheostomy after an average time interval of 1.9 years (range 0.5–3.5 years). Tracheostomy was performed in 80% of referred patients with CP older than 10 years, while surgical intervention was uncommon in children younger than 5 years. There was a statistically significant correlation between the age of the children and the performance of a tracheostomy (Pearson's correlation coefficient 0.68, $p = 0.005$).
Conclusions: The severity of the airway obstruction in children with CP tends to increase with age. We postulate that this increase results from worsening hypotonia of pharyngeal musculature. Children with CP and severe upper airway obstruction are likely to require tracheostomy as they grow older.

1. Introduction

Cerebral palsy (CP) is a non-progressive disorder of motor function with prevalence of approximately 1:500 live births [1]. Apart from learning difficulties, motor and speech impairment, children with CP are at high risk of developing severe airway obstruction either in form of obstructive sleep apnea [2–4] or even as awake upper airway obstruction [5]. Although poor neuromuscular tone is the main underlying etiology, maxillary hypoplasia, retrognathia and glossoptosis, laryngomalacia, seizures, gastro-esophageal reflux and increased oropharyngeal secretions have also been associated with the airway problems [5,6]. The contribution of many factors to the airway obstruction and the presence of multiple additional co-morbidities in children with CP make their management very challenging.

In order to treat the airway obstruction in children with CP, several interventions have been suggested depending on clinical (signs of respiratory distress, recurrent airway infections or obstructive sleep apnea) and laboratory (overnight oxymetry, polysomnogrphy) findings [2–8]. Continuous or biphasic positive airway pressure (CPAP and BIPAP respectively) represents a non-invasive approach to these children [7]. Adenotonsillectomy is widely recommended as the primary surgical procedure with promising results in many cases [2,6–8]. Depending on the underlying cause, septoplasty, turbinectomy uvulopalatopharyngoplasty may also help to improve the airway symptoms [7,8]. For more severe and persisting symptoms in patients with additional facial dysmorphic features, even more aggressive and complex techniques such as floor of mouth degloving, tongue hyoid advancement and tongue reduction, mandibular distraction and mandibular osteotomies have been described [8–10]. Finally, given the complexity and severity of cases with CP, tracheostomy, the traditional method of dealing with upper airway obstruction when other measures fail, may still be required [1,6,7].

Although the airway problems in patients with CP have been described, most of the limited previous works have focused on the surgical management. Studies on the initial presentation and the progress of the airway obstruction over time are unknown to the authors. The objective of the present study was to describe the

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natural history of upper airway obstruction in pediatric patients with CP and identify the expected timing for the different interventions.

2. Materials and methods

2.1. Basic settings and patients selection

This retrospective chart review was carried out in a tertiary, referral Pediatric Otolaryngology department. Approval was obtained by the Clinical Audit department.

The medical notes of patients younger than 16 years with CP, who have been referred to our department from January 2006 to January 2012 with upper airway obstruction symptoms, were retrospectively reviewed. The presenting symptoms and signs following detailed history and otolaryngological clinical examination involved snoring, inspiratory stridor, sleep apnea episodes witnessed by the caregivers or diagnosed by overnight oxymetry or polysomnography with or without the need of nocturnal oxygen supply and sternal or sub/intracostal recessions. In order to identify all patients with CP and airway obstruction the medical notes were reviewed with the help of the Clinical Audit department and the coding system used for the outpatient department, the inpatient admissions and the theater documentation.

2.2. Examined factors

The age at the initial presentation and the gender were recorded. In all cases, whether conservative or surgical, treatments were documented. Additionally, the age at the time of every surgical intervention and the additional co-morbidities were also noted. Conservative treatment included nocturnal oxygen supply or CPAP/BIPAP, while the usually performed surgeries in such patients in our department involved adenotonsillectomy, upper airway endoscopy (rigid pharyngo-laryngo-tracheoscopy), supraglottoplasty and tracheostomy. Pharyngo-laryngo-tracheoscopy was performed in all children to assess their upper airway. In children undergoing tracheostomy after failure of the initially performed surgical intervention, the time period between the primary surgery and the tracheostomy was documented.

Surgical intervention was proposed when conservative management with CPAP/BIPAP has failed. In particular, children with CP who had signs of respiratory distress and clinical symptoms or laboratory findings of obstructive sleep apnea, despite their overnight ventilation were offered further surgical treatment. In some children with recurrent airway infections upper airway endoscopy was offered, mainly for diagnostic reasons. The decision for surgical treatment was met following thorough consultation of the caregivers.

Adenoidectomy was performed with either the traditional technique with the St. Claire Thompson's curettes or with the monopolar suction (38 W). Tonsillectomy was performed using bipolar dissection technique (5–7 W). Supraglottoplasty was performed endoscopically or microscopically using cold dissection technique for excising the aryepiglotic folds and reducing the arytenoid redundant tissue; the intra-arytenoid area was kept in all cases intact.

All patients were admitted to the pediatric high dependency or intensive care unit following surgical intervention. Finally, the follow-up period for all patients was documented.

Children with CP who were admitted to our department and treated for other otolaryngological problems, non-related to upper airway obstruction were excluded from the present study.

2.3. Statistical evaluation

Pearson's correlation was used to identify any linear correlation between the age of the patient and the performance of tracheostomy (SPSS V.21). $p < 0.05$ was considered statistically significant.

3. Results

3.1. Patients

Fifteen children with CP and upper airway obstruction symptoms were admitted to our department, seven girls and eight boys. The average age at the time of presentation was 8 years (range 3–13.3 years) and the mean follow-up time 2.6 years (range 0.5–6.5 years). All children had symptoms of upper airway obstruction and had all been previously treated with nocturnal or/and daytime oxygen. They all had additional co-morbidities varying from gastroesophageal reflux to global development delay; however, major dysmorphisms were not observed (Table 1).

3.2. Interventions

At the time of the last evaluation three children did not need any surgical interventions as they had all responded satisfactorily to oxygen supply and CPAP. Adenotonsillectomy was performed in 11 out of 15 cases (73.3%), supraglottoplasty in three (20%, patients 1, 8 and 9) and tracheostomy in eight patients (53.5%). The mean age for adenotonsillectomy was 9.1 years (range 4.5–14 years), while the average age at the time of tracheostomy was 11.6 years (range 7.5–15 years). Seven out of the 11 children (63.6%) initially treated with adenotonsillectomy, had to undergo tracheostomy

Table 1
Demographic data and morbidities in the CP children.

Patient	Gender	Age at referral (years)	Morbidities
1	Female	4	Prematurity, OSA
2	Female	13.3	Multicystic encephalopathy, OSA, GORD
3	Male	11.5	Chronic respiratory disease, visual impairment, epilepsy, OSA
4	Male	11	Scoliosis, visual impairment, OSA
5	Male	8.3	Epilepsy, OSA, increased pharyngeal secretions
6	Female	7.7	OSA, increased pharyngeal secretions, GORD
7	Male	3	Tracheomalacia, OSA
8	Female	4.2	OSA, visual impairment
9	Female	4.2	OSA, visual impairment
10	Male	10.2	Epilepsy, visual impairment, OSA, increased pharyngeal secretions
11	Male	10	OSA, increased pharyngeal secretions
12	Male	7.3	Epilepsy, GORD, OSA
13	Male	9	GORD, OSA, epilepsy
14	Female	9.7	OSA, visual impairment, increased pharyngeal secretions
15	Female	6	GORD, OSA, increased pharyngeal secretions, epilepsy

[a] OSA: obstructive sleep apnea and GORD: gastroesophageal reflux disease.