Tonsillectomy and adenoidectomy in patients with Down syndrome

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Abstract

Tonsillectomy and adenoidectomy in patients with Down syndrome usually relieves symptoms of upper airway obstruction due to adenotonsillar hypertrophy. A retrospective review of 16 patients with Down syndrome who underwent a tonsillectomy and adenoidectomy was conducted to define early postoperative morbidity, and to evaluate the results of the procedure. Fifteen of 16 patients had upper airway obstruction manifest by adenotonsillar hypertrophy. One had adenoid hypertrophy only. Tonsillectomy and adenoidectomy (13), adenoidectomy (1), tonsillectomy (1), and UP3/adenoidectomy (1) were performed on these patients. Four patients (25%) required observation in an intensive care setting the first night. An average hospital stay of 2.1 days was noted with a range of 0 to 7 days. Significant postoperative apnea was common, and oxygen was used in over 60% of patients. Symptoms were resolved in 69% of patients at last follow up. In conclusion, tonsillectomy and adenoidectomy can be a useful procedure for children with Down syndrome and obstructive sleep apnea, however overnight hospitalization for observation and treatment of persistent apnea is appropriate.

Keywords: Tonsillectomy; Adenoidectomy; Down syndrome; Complications; Airway obstruction

1. Introduction

Children with Down syndrome are at increased risk for obstructive sleep apnea secondary to tonsil and adenoid hypertrophy. Relatively few articles have been published on children with Down syndrome who have undergone a tonsillectomy

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and adenoidectomy [1–8]. Most articles have evaluated the results of tonsillectomy and adenoidectomy to cure obstructive symptoms. Very little has been written about the early post operative morbidity of procedures in these children, a problem of significant concern in the present medical climate of cost savings in which more and more procedures are being done on an outpatient basis.

Down syndrome occurs in approximately 1.5 of 1000 births and accounts for 10% of mentally retarded persons [6,9,10]. Children with Down syndrome commonly present with otolaryngologic problems including frequent upper respiratory infections (61% of patients), chronic otitis media, small ear canals, hearing loss (70–80% of patients), and hypothyroidism [4]. Children with Down syndrome also fall into the group of children with craniofacial anomalies and neurologic abnormalities which predispose them to obstructive sleep apnea [10–12]. Children with Down syndrome are at increased risk for obstructive sleep apnea due to their relatively small mid-face and cranium [13], their relatively narrow nasopharynx [9], their relatively large tongue [14], muscular hypotonia [15], obesity [7], increased susceptibility to upper respiratory infections [3] and relatively small larynx [1,2,6,16,17]. Up to 50% of children with Down syndrome may have congenital heart disease which potentially predisposes these children to cor pulmonale, a known complication of prolonged sleep apnea [18–20]. Because of these factors, the incidence of obstructive sleep apnea in patients with Down syndrome has been estimated at from 54% to 100% of patients [2,4,7].

Because of this high incidence of obstructive sleep apnea in patients with Down syndrome, the majority of patients are considered for surgical management somewhere during their course of treatment. Tonsillectomy and adenoidectomy continue to be the most commonly performed major surgical procedures on children in the United States [21], and obstructive sleep apnea in children has become the most common reason for tonsillectomy and adenoidectomy [22–24]. Strome in 1981 reported one of the first series of patients with Down syndrome undergoing tonsillectomy and/or adenoidectomy [9]. They noted benefit in only 3 of the 16 patients.

Strome in 1986 however reported uniform success in relieving obstructive sleep apnea in 5 patients undergoing tonsillectomy, adenoidectomy and uvulopalatopharyngoplasty (UP3) [8]. Similarly, improvement in symptoms such as apnea (83%), and snoring (81%), was noted in 21 patients in another series [4]. Even in severe cases, Kasian in 1987 reported that 2 of 3 patients obtained relief from pulmonary hypertension due to OSA after undergoing a tonsillectomy and adenoidectomy [25]. Donaldson in 1988 and Hultcrantz in 1991 reported reasonable success with oropharyngeal surgery in patients with Down syndrome [1,6]. Finally in 1991, 8 children were reviewed with success in the majority but complete normalization in only three [7].

While tonsillectomy and adenoidectomy improves obstructive symptoms at least in some patients with Down syndrome, the perioperative risks of the surgery have not been systematically reviewed. Due to the underlying cardiac disease, craniofacial anomalies, hypotonia, and other medical problems, an increase in perioperative risks would be expected in these patients. While outpatient tonsillectomy and adenoidectomy has been shown to be safe in most patients, it is not reasonable to assume that this procedure is safe on an outpatient basis for all subgroups of patients.
It was therefore felt appropriate to evaluate our experience with patients with Down syndrome who underwent tonsillectomy and/or adenoidectomy not only to evaluate the effectiveness of the procedure, but also to evaluate the need for early post operative observation and/or intervention for complications.

2. Materials and methods

A retrospective review of patients with a diagnosis of Down syndrome who underwent tonsillectomy and/or adenoidectomy was performed. Patients were identified by reviewing inpatient and OR records from January 1986 to December 1993. All charts were reviewed and data collected. Descriptive statistics only were utilized.

3. Results

Sixteen patients were identified over a 7-year period who had Down syndrome and also underwent a tonsillectomy and/or adenoidectomy (Table 1). These patients were identified from a clinic which evaluates over 9000 patients per year.

Preoperative symptoms included snoring 63%, witnessed apnea 38%, mouth breathing 31%, restless sleep 13%, and recurrent tonsillitis 6%. Tonsil size was graded on a 0 to 4+ scale, 0 being absent, 1+ filling less than 25% of the oropharynx, up to 4+, filling greater than 75% of the oropharynx. Preoperative signs included 2+ tonsils in 14%, and 3+ or larger tonsils in 86%. A large tongue was specifically noted and documented in 25% of the patients. Other findings included small nares, a small midface, and a short, thick neck.

Preoperative testing included in house pulse oximetry in 4 patients with documented desaturations below 75% in all and as low as 30% in 1. Only 1 patient had a pre-op polysomnogram which revealed an apnea/hypopnea index of 5.5 (i.e. the number of apneas of greater than 10 s duration, and the number of hypopneic spells (reduction of tidal volume with desaturation of > 5%) per hour of sleep).

Obstructive sleep apnea was diagnosed based on a history of snoring, frequent witnessed apneic spells, restless sleep, and/or daytime somnolence, with corresponding physical findings. A polysomnogram, or overnight pulse oximetry was obtained when diagnosis was uncertain. Fifteen of the 16 patients were diagnosed with adenotonsillar hypertrophy with obstructive sleep apnea based on history and physical findings. One patient was felt to have adenoid hypertrophy alone.

Table 1
Demographic data of the 16 patients evaluated

<table>
<thead>
<tr>
<th>Total patients</th>
<th>16</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>56% (9)</td>
</tr>
<tr>
<td>Female</td>
<td>44% (7)</td>
</tr>
<tr>
<td>Caucasian</td>
<td>81% (13)</td>
</tr>
<tr>
<td>Black</td>
<td>19% (3)</td>
</tr>
</tbody>
</table>
PROCEDURES

Fig. 1. Thirteen patients underwent tonsillectomy and adenoidectomy (T/A), 1 tonsillectomy (T), 1 adenoidectomy (A), and 1 adenoidectomy and uvulopalatopharyngoplasty (A/UP3).

Concomitant cardiac disease was quite common occurring in 11 of 16 (70%) of our patients. Concomitant otitis media was noted in 63%.

Thirteen patients underwent tonsillectomy and adenoidectomy only, 1 underwent a tonsillectomy, 1 underwent an adenoidectomy, and 1 underwent an adenoidectomy with UP3 (Fig. 1). Electrocautery tonsillectomy, and curette adenoidectomy were performed by both ENT residents and staff. Nine patients underwent myringotomy and PE tube placement at the time of their surgery. The average age at the time of surgery was 4.9 years with a minimum of 1 year and a maximum of 14 years.

Two patients (age 3 and 3.3 years) were discharged home the day of surgery tolerating an oral diet well, and not demonstrating any evidence of desaturations, or airway obstruction. 75% of the remaining patients were admitted to the floor where they were observed on a pulse oximeter. 25% or 4 of the remaining patients required admission to the intensive care unit. These patients were admitted to the ICU for documented severe upper airway obstruction and desaturations in the post anesthetic care unit. The average duration of hospitalization was 2.1 days with a range of 0–7 days. No patients required intubation post operatively; however, one required a nasopharyngeal airway the first post operative night for severe upper airway obstruction and apnea.

Oxygen was used in 10 of the 16 patients with 63% of patients requiring a 40% oxygen hood to maintain oxygen saturation above 90%. The lowest post operative saturations based on pulse oximetry over the first night averaged 63%. Steroids were used in 2 (13%) patients while antibiotics were recommended in all patients. Post op-
RESULTS OF TREATMENT

Fig. 2. Surgical treatment for OSA in patients with Down syndrome resulted in significant improvement in 11 patients (69%), no change (same) in 4 patients (25%), and persistent OSA (trach) in 1 patient (6%).

Operative complications were noted in 6 patients. One patient developed stridor and significant retractions with desaturations post operatively and required monitoring in the ICU. Another patient developed pulmonary edema in the early post operative period. One patient had witnessed apnea with desaturations and also vomiting. One patient was observed for bleeding which did not require operative intervention. Prolonged dysphagia was noted in 1 patient and fever in another.

Eleven of 16 patients (69%) were subjectively improved or their symptoms were resolved with treatment (Fig. 2). This meant patients had no further snoring, witnessed apneic spells, or desaturations based on pulse oximetry. One patient failed approximately 7 months after his initial procedure when he developed respiratory syncytial virus pneumonitis and required intubation followed by tracheotomy. He still requires a tracheotomy for PSG documented OSA nearly 1 year later.

4. Discussion

Obstructive sleep apnea is a common problem in children with Down syndrome. Along with a general medical evaluation, these patients should be evaluated for obstructive sleep apnea as early as 5–6 months of age and on a routine basis thereafter [1]. Routine evaluation should include a history for significant obstructive sleep apnea manifest by snoring, mouth breathing, restless sleep, and witnessed apneic
spells. Parental history is the most reliable indicator in documenting obstructive
sleep apnea in children [8]. In particular snoring in all positions is the sine qua non
of OSA. Overnight pulse oximetry can also be helpful in borderline situations. The
diagnostic test which provides the most information is polysomnography (PSG).
While this provides a very complete representation of sleep and thoroughly differenti-
tiates between obstructive and central events, PSG in patients with Down syndrome
may be somewhat difficult to perform. PSG may be difficult to administer due to
the mental status of the patient [1], the test may disturb normal sleep patterns [6],
and because of this PSG may not accurately reflect the degree of sleep disturbance.
Therefore, clinical correlation is very important in determining severity of disease
and deciding on treatment. While many of the patients in this retrospective study
were not evaluated by PSG, our routine now is to obtain at least a post operative
PSG 6 weeks after surgery, and usually both a pre and post op study, to evaluate
these patients more objectively.

Once a decision has been made to operate, and prior to operative intervention,
patients with Down syndrome require evaluation of thyroid function, C-spine
stability, and cardiac status. Thyroid functions should be performed on all patients
as hypothyroidism is relatively common [1]. Evaluation of the cervical spine for
atlanto-axial instability is important as this problem has been noted in up to 10–20%
of children with Down syndrome [4]. Fully 50% of patients with Down syndrome
have congenital heart disease, and surgical intervention could result in significant
cardiac compromise in undiagnosed cases [17]. Some authors recommend that bron-
choscopy be performed on all patients at the time of tonsillectomy and adenoidecto-
my as 5 out of 5 patients in one series were found to have anomalies of the larynx,
trachea, or proximal bronchi [8].

Prior to deciding whether to treat patients with obstructive sleep apnea surgically,
acute infections should be treated and cardiopulmonary disease should be addressed.
In an acute situation, a nasal pharyngeal airway may be helpful and steroids may
help reduce not only nasal edema but also enlargement of the tonsils and adenoids
[6].

Surgical treatment for patients with obstructive sleep apnea and Down syndrome
should be individualized. In our study, tonsillectomy and adenoidectomy was suf-
ficient for the majority of our patients. Hultcrantz also felt that this procedure was
sufficient in the majority of patients [1]. UP3 is another recommended procedure,
and intraoperative assessment of the importance of the palate and the uvula in ob-
struction with possible extension of the procedure is reasonable. Strome et al. en-
couraged UP3 for all patients as their results with this extended procedure was
significantly better than for tonsillectomy alone [8]. They also found that the majori-
ty of patients did not have significantly enlarged adenoids, and that an adenoidecto-
my should only be added when obstructing adenoids are noted. Macroglossia is
quite common in children with Down syndrome. In situations in which the tongue
is obviously obstructing/protruding from the mouth, a tongue reduction may be
helpful. Unfortunately there is little documented on the effectiveness of this very in-
vasive procedure [6,8]. Tracheotomy is the definitive procedure for upper airway ob-
struction in children. Post operative care can be complicated by poor patient
compliance, frequent infections, and difficult care [1,8]. We have not recommended tracheotomy unless more conservative operations have failed. In summary, surgical care should be recommended based on the physical examination, sleep study results, intraoperative evaluation, and the desires of the parents. While tonsillectomy and adenoidectomy is adequate in many situations, more aggressive treatment such as UP3, tongue reduction, and tracheotomy may be necessary.

Post operative care in patients with Down syndrome after tonsillectomy and adenoidectomy can be crucial to the patient’s safety. Swallowing is often delayed and hospitalization is prolonged [8]. Some authors feel that resurfacing by approximating the tonsil pillars, and the use of steroids can improve postoperative outcome with early return of swallowing, and discharge home within 24–36 h. Patients should be admitted post operatively as persistent obstructive apnea and desaturations are quite common, occurring in over 50% of our patients. Children should be admitted to the intensive care unit if significant apnea is witnessed in the recovery area. Hospitalization up to 1 week postoperatively is necessary in some patients. The use of nasal trumpets, intubation or other form of artificial airway is occasionally necessary (<10%). Oxygen supplementation is often required and pulse oximetry monitoring is recommended for all patients.

Complications after tonsillectomy and adenoidectomy include persistent obstructive apnea with desaturations which often requires oxygen and may require more invasive treatment. Fever is a relatively common problem after tonsillectomy and adenoidectomy. Nausea and vomiting may also occur. Bleeding is probably no more common in these children than that experienced in other patients. Pulmonary edema which is usually post obstructive in nature can also occur and may be related to underlying cardiac disease. Patients with Down syndrome may be at increased risk for velopharyngeal insufficiency due to their hypotonia [4]. VPI may be more common after UP3, however this has not been studied in detail. Finally, nasopharyngeal stenosis may occur postoperatively, and may be more common after UP3, especially if done with an adenoidectomy.

Conclusions

1. Tonsillectomy and adenoidectomy is successful in relieving obstructive sleep apnea in the majority of patients with Down syndrome (69%). More aggressive intervention such as UPPP, C-Pap, tongue reduction, or tracheotomy are necessary in some patients.
2. Preoperative evaluation should include assessment for cardiac, thyroid, and cervical abnormalities.
3. Surgical planning should be based on the severity of disease and the physical findings. Initially conservative procedures such as T and A are reasonable.
4. Follow up sleep studies are indicated to evaluate for the need for more aggressive treatment in patients with persistent symptoms. Optimally, pre and postoperative PSG should be obtained to evaluate these patients objectively.
5. Patients with Down syndrome should be admitted to the hospital post operatively as persistent obstructive sleep apnea and other complications are common (63%).
Intensive care unit monitoring is often necessary (25%). Post operative hospitalization may be prolonged.

References