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ORIGINAL ARTICLE

Effects of adenotonsillectomy in children with Down Syndrome

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ABSTRACT

BACKGROUND: Down Syndrome (DS) is one of the most frequent genetic abnormalities and the related otolaryngologic disorders may affect a full potential development and the quality of life of these patients. The aim of the present study was to evaluate which treatment, medical or surgical, may improve otolaryngologic symptoms.
METHODS: We conducted a case-control study considering the number of acute pharyngo-tonsillitis febrile episodes (PTFE), presence of obstructive sleep apnea (OSA) and middle ear disease in patients with DS referred to our centers. We selected 34 subjects with more severe symptoms and clinical course subdivided in two groups, the first group constituted by patients that underwent conservative therapy and the second group by those who underwent surgical therapy.
RESULTS: After surgical treatment we found a significant ($P<0.01$) reduction of PTFE and OSA grade with a normalization of tympanometry ($P<0.01$). Conversely, we observed a reduction of symptoms only in few cases ($P>0.05$) that underwent medical therapy. No major complications related to DS were observed in surgical therapy.
CONCLUSIONS: Adenotonsillectomy appears to be more helpful than medical therapy in DS subjects determining OSA, balanced with surgical risks/benefits, respiratory and ear function improvement.

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KEY WORDS: Down Syndrome - Middle ear - Obstructive sleep apnea - Tonsillectomy - Adenoidectomy.

Down Syndrome (DS) is the commonest genetic abnormality and affects approximately 1:660 to 1:800 children born.[1] In the last years advances in surgical treatments of congenital heart defects have greatly enhanced the survival of DS, but the increasing life expectancy has revealed the presence of unexpected pathological processes.[2]

It is widely accepted that DS children suffer from more frequent infections than normal children, and most studies agree that these affect mainly the respiratory tract.[3] Patients with DS have several morphologic abnormalities (mid-face hypoplasia, Eustachian tube dysfunction, abnormalities of the oropharynx and rhinopharynx, and shortened palate) that predispose them to ear, nose, and throat (ENT) problems. Common ENT manifestations in DS children include ear infections and hearing loss, obstructive sleep apnea (OSA) subglottic stenosis, chronic rhinorrhea and sinusitis, as well as important anesthetic considerations.[4]

Approximately one-third of children with DS have recurrent otitis media with effusion (OME) that requires close monitoring.[5] Etiologic factors include an increased incidence of upper respiratory tract infections (URTI), possibly due to the immaturity of the immune system. In addition, the mid-face hypoplasia, the shape of Eustachian tube, more cylindrical and smaller in width, and the hypotonia of the tensor veli palatini muscle predispose to chronic ear disease.[4]

All the mentioned ENT disorders can affect the potential development and quality of life of these patients. Consequently, appropriate treatments may be required to manage the disease causing ENT manifestations. For instance, hearing loss can affect a child's educational, language, and social development and then it must be diagnosed and treated aggressively.

Furthermore, the prevalence of OSA is greater among children with DS or other chronic pediatric or psychiatric conditions, than among children in general. Good-quality sleep is important for learning, attention and memory processes.[6] On the other hand, sleep disruption may exacerbate learning difficulties and disturbed behavior that are the consequence of the developmental disorder itself.[7] OSA could be related also to sensorineural hearing impairment.[8]

The adenotonsillectomy could be one of more reliable surgical solution to ENT symptoms (*i.e.*, otitis due to Eustachian tube obstruction, OSA due to tonsil hypertrophy); even if is a documented treatment for OSA in non-syndromic children[9] there is actually divergence in literature supporting adenotonsillectomy for OSA in DS children.[10-13]

The aim of the present case-control study was to evaluate the efficacy of adenotonsillectomy in children with DS.

Materials and methods

A case-control study of DS patients observed and/or treated in our Institutions was done. The patient personal data were collected and recorded.

TABLE I.—*Parameters evaluated in the first clinical examination.*

Evaluated parameters
Number of pharyngo-tonsillar febrile episodes
Increase in body temperature ≥38 °C with catarrhal or follicular angina with reference to the past 12 months. We considered three classes according to the frequency of febrile episodes: • Class 0: no or 1 febrile episode per year; • Class I: 2-4 episodes per year; • Class II: 5 or more episodes per year
Tonsillar hypertrophy
Clinical evaluation of the degree of tonsillar hypertrophy performed according to the Mallampati criteria: Grade 0: tonsils within tonsillar fossa; Grade 1+: tonsils <25% of space between pillar; Grade 2+: tonsils <50% of space between pillars; Grade 3+: tonsils <75% of space between pillars; Grade 4+: tonsils >75% of space between pillars
Endoscopic evaluation of the nose and rhinopharynx
Performed with the patient in a sitting and lying position with a 2.7-mm flexible endoscope by the same investigator. No decongestant was used.
Nasal status was scored as follows: (0) normal turbinates/no discharge; (1) clear and thin discharge; (2) thick and purulent discharge; absence (0) and presence (1) of edema.
Adenoid hypertrophy was scored: (1) choanal openings free or adenoid tissue confined to the upper segment in the rhinopharynx (<25%); (2) adenoid tissue occupying only the upper segment in the rhinopharynx (≥25% to <50%); (3) adenoid tissue extended over the half volume of rhinopharynx (≥50% to <75%), (4) total choanal obstruction (>75% to 100%)
Overnight polysomnography (PSG) (Apnoescreen II plus)
Record of the respiratory distress index (RDI), the apnea-hypopnea index (AHI) and the mean oxygen saturation (SaO_2%) during sleep. OSA was defined absent (RDI 1-3, SaO_2 97-100%), mild (RDI 3-5, SaO_2 96%), moderate (RDI 5-10, SaO_2 95%) and severe (RDI>10, SaO_2 94%)
Middle ear involvement
Diagnosed on the basis of otoscopy (tympanic atelectasis), tympanometry (shape of curve type A, B, C) and acoustic reflectometry, to verify the presence of middle ear disease

TABLE II.—*Characteristics of Down Syndrome patients.*

Age (years)	N.	Obstructive sleep apnea				Middle ear disease	Pharyngotonsillar febrile episode		
		Absent	Mild	Moderate	Severe		Class 0	Class I	Class II
0-1	30	2 (6.6%)	5 (27%)	8 (27%)	15 (50%)	22 (73.3%)	20 (66.6%)	6 (20%)	4 (13%)
1-2	29	5 (17%)	5 (17%)	5 (17%)	14 (48%)	20 (68.9%)	8 (27.5%)	13 (44.8%)	8 (27.5%)
2-3	18	3 (17%)	1 (5.5%)	9 (50%)	5 (28%)	13 (72.2%)	5 (27.5%)	8 (44.4%)	5 (27.7%)
3-4	19	5 (26%)	3 (5.2%)	7 (36%)	6 (31%)	10 (52.6%)	4 (21%)	7 (36.8%)	8 (42.1%)
4-5	25	9 (36%)	5 (10%)	6 (24%)	5 (20%)	12 (48%)	10 (20%)	10 (40%)	5 (20%)
5-6	27	7 (26%)	7 (26%)	8 (30%)	5 (18%)	12 (44.4%)	8 (29.6%)	13 (48.1%)	6 (22.2%)
Total	148	31 (21%)	24 (16%)	43 (29%)	50 (34%)	89	55 (37.2%)	57 (38.5%)	36 (24.3%)

TABLE III.—*Selected patients (N.=34) characteristics before and after surgical and medical treatment.*

Type of treatment	Before				
	PTFE Class I	PTFE Class II	Moderate OSA	Severe OSA	MEI
Surgical treatment	9/17 (52.9%)	8 /17 (47%)	10/17 (58.8%)	7/17 (41.1%)	17/17 (100%)
Medical treatment	9/17 (52.9%)	8/17 (47%)	10/17 (58.8%)	7/17 (41.1%)	17/17 (100%)

PTFE: pharyngotonsillar febrile episode; OSA: obstructive sleep apnea; MEI: middle ear involvement.

The Institutional Ethics Committee approved the study protocol. All parents gave written informed consent for their children's participation in the clinical pathway and in the study. All subjects were accompanied during the study by one or both parents.

During the first clinical consultation patients were evaluated for: number of pharyngo-tonsillar febrile episodes (PTFE) referred to the past 12 months, tonsillar hypertrophy grading, endoscopic evaluation of the nose and rhinopharynx with grading of obstruction, overnight polysomnography (PSG), and middle ear involvement (otoscopy, tympanometry, stapedial reflex). Parameters definitions are reported in Table I.

A group of patients with severe ENT manifestations, matching the inclusion criteria, was addressed for treatment.

Severe ENT involvement was defined by the simultaneously presence of: number of pharyngo-tonsillar febrile episodes (PTFE) class I and II; tonsillar hypertrophy 3 to 4; adenoid hypertrophy grade 3 to 4; PSG: OSA moderate to severe; bilateral pathological tympanograms as flat (type B) or peak compliance below zero (type C).

Exclusion criteria: age under 2 years, severe contraindication to surgery due to cardiac malformations, pulmonary issue.

We subdivided the group of patients with more severe ENT involvement into two subgroups by a blocked randomization (size of block 4 patients). The first subgroup includes those underwent cold adenoidectomy and cold dissection tonsillectomy with bipolar coagulation when needed. The surgery was performed in Rose position under general anesthesia with oral-tracheal intubation. All patients were hospitalized overnight after surgery. The complications occurred were recorded. The second subgroup included those underwent medical therapy with beclomethasone 200 μg twice a day and N-acetylcysteine 200 mg/twice/day by aerosol for two weeks; amoxicillin with clavulanate 25 mg/kg twice per day were administered only in case of recurrence of tonsillar symptoms or exacerbation of nasal infections.

Both groups were followed-up with a clinical control every 3 months for 18 months with objective examination, nasal endoscopy, otoscopy, tympanometry and acoustic reflectometry. A PSG was repeater after 12 months.

Statistical analysis

Distributions of continuous variables in different groups were analyzed by Student's *t*-test parametric method. For categorical variables the comparisons were performed by using the χ^2 test in MS Excel.

Results

A total of 148 DS patients were observed in our Institutions among 2004-2010. Mean age was 3.4 years. The 54.7% of patients was male. Clinical characteristics of patients are reported in Table II.

After the first clinical consultation we selected 34 patients matching our criteria with more severe ENT involvement; mean age 5,1 years old (range 3-6) for surgical subgroup and 5.3 years old (range 3-6) for medical subgroup (P>0.05).

After 18 months of follow-up in the subgroup of patients who underwent adenotonsillectomy there was a statistically significant reduction of number of PTFE class I and II (from 17/17 to 2/17 patients, P<0.01), of moderate and severe OSA (from 17/17 to 3/17 patients, P<0.01) and of pathological tympanograms from (from 17/17 to 2/17 patients, P<0.01).

No complications occurred after adenotonsillectomy.

Conversely, in the subgroup of patients who underwent medical therapy there was no statistically significant reduction of number of PTFE (from 17/17 patients to 14/17, P>0.05), of moderate and severe OSA (from 17/17 patients to 15/17, P>0.05) and of pathological tympanograms (from 17/17 patients to 14/17, P>0.05).

Overall statistical significance showed a P<0.01 between surgical and medical treatment.

All results are summarized in Table III and IV.

Discussion

Health advances in medical care on the one hand have lengthened the life expectancy for children with DS, on

At 18 months				
PTFE Class I	PTFE Class II	Moderate OSA	Severe OSA	MEI
2/17 (11.7%)	0/17	3/17 (17.6%)	0/17	2/17 (11.7%)
8/17 (47%)	6/17 (35.3%)	10/17 (58.8%)	5/17 (29.4%)	14/17 (82.3%)

TABLE IV.—*Medical* versus *surgical treatment results after 18 months of follow-up.*

Parameter	Surgical treatment			Medical treatment			Overall P value
	Before	After	P value	Before	After	P value	
PTFE Class I, II	17/17	2/17 (11.7%)	<0.01	17/17	14/17 (82.3%)	>0.05	<0.01
Moderate. severe OSA	17/17	3/17 (17.6%)	<0.01	17/17	15/17 (88.2%)	>0.05	<0.01
FT	17/17	2/17 (11.7%)	<0.01	17/17	14/17 (82.3%)	>0.05	<0.01

the other hand increased the prevalence of morbidity. For this reason, because of their enhanced predisposition for the development of ENT manifestations, DS population should have particular attention with regards to screening for the onset of ENT problems and their treatment.

Common ENT manifestations in children with DS include: increased incidence of middle ear infections and conductive hearing loss, OSA, chronic rhinorrhea and rhinosinusitis. The DS patients have moreover increased anesthetic risk. OSA, if untreated, could cause serious pulmonary sequelae such as hypercarbia, acidosis, and pulmonary hypertension with possible cor pulmonale. However, long-term sequelae of OSA, as well as chronic middle ear infections can be avoided with early diagnosis and proper management.[4, 14] Therefore, to improve the quality of life in DS children the clinician should be familiar with different presentation of ENT manifestations including hearing loss, sleep-disordered breathing issue, as well as other health conditions like hypothyroidism, atlanto-axial instability and cardiac diseases, which can potentially affect the management of the patient.[15]

As showed by our analysis the patients underwent adenotonsillectomy had a significant reduction of PTFE and OSA symptoms, and a normalization (type A) of timpanometry (Table IV). Similarly, Kavanagh reports an improvement in OSA symptoms in 17 cases but some other authors report no significant functional advantages in surgical treatment and suggest a limited use of it.[16, 17] Only in a limited number of cases of the group that was managed with medical therapy, we observed a reduction of PTFE and an improvement of OSA and timpanometry (Table IV). Our data suggest that the surgical treatment was useful in recovering respiratory obstruction and middle ear function, and consequentially, in rehabilitation and social integration.[18] Choose of the medical therapy adopted: aerosol beclometasone instead of steroids nasal spray was linked to reduce costs for the parents preventing the abandoning of the study.[19]

In our opinion, as showed also in previous report in non-syndromic adults,[9] the tonsillectomy improves nocturnal respiratory parameters and daytime symptoms, as well as cardiovascular complications. However, despite its effectiveness, the surgical treatment may expose the DS patients to anesthetic complications such as bradycardia, airway issue, difficult intubation, and post-intubation croup.[4] For these reasons it is important that the anesthesiologist has a good understanding of the pathophysiology of DS children. Thus overnight hospitalization after surgery and special vigilance in the postoperative period should be needed.[5] No complications linked to surgical procedures, or general anesthesia, were recorded in our series.

DS subjects are predisposed to a higher functional respiratory vulnerability and 70% of children with DS show residual or recurrent OSA after surgery. On account of this, we recommend a careful clinical and instrumental follow-up extended also after following years.

Conclusions

The adenotonsillectomy appears to be more helpful than medical therapy in DS subjects with OSA, improving significantly PTFE, respiratory and middle ear function. Because of high risk of anesthetic and postoperative complications the candidates for surgery should be only children with high frequency and severity of febrile episodes, pathological tympanometry as well as children with high degree of adenotonsillar hypertrophy with a moderate to severe OSA influencing their quality of life and psychological condition.

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Conflicts of interest.—The authors certify that there is no conflict of interest with any financial organization regarding the material discussed in the manuscript.

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