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The Importance and Place of Adenotonsillectomy in Syndromic Children

Original Investigation

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Abstract

Objective: Upper airway obstruction and sleeping disorders are important issues in syndromic children, including mouth, lower-upper jaw, or all facial abnormalities. Tonsillectomy and/or adenoidectomy is required because of systemic problems and upper airway obstruction that increase the existing systemic problems, except those anomalies. However, tonsillectomy and/or adenoidectomy are mostly avoided because of the tendency to cause both intubation/perioperative systemic problems and respiratory complications in the postoperative period and in delays in the oral intake. However, these surgeries are sometimes required. In this context, we present our experience related with performing tonsillectomy and/or adenoidectomy in syndromic children admitted to our hospital.

Methods: We retrospectively examined the data on tonsillectomy and/or adenoidectomy performed in syndromic patients in our clinic between 2001 and 2011.

Results: We did not observe any postoperative complications in adenoidectomy and/or tonsillectomy performed by the same surgeon in 14 syndromic cases.

Conclusion: It should be noted that respiratory problems may arise from many different anatomical regions in syndromic patients. Therefore, surgery should be performed taking into consideration all of these factors in these patients. These patients must be hospitalized in the postoperative period.

Keywords: Adenoidectomy, tonsillectomy, syndromic disease, complication

Introduction

Tonsillectomy and/or adenoidectomy surgeries are frequently performed because of frequent upper respiratory tract infections or respiratory disorders during sleep in children. Obstructive sleep apnea is more common in children with craniofacial anomalies than in healthy children (1). Obstructive sleep apnea can be more serious in syndromes that have symptoms such as craniosynostosis, maxillo-mandibular hypoplasia, midfacial developmental defects, micrognathia, and muscular hypotonia (Down syndrome, syndromic craniosynostosis, cerebral palsy, Prader–Willi syndrome, Apert syndrome, Crouzon syndrome, Pfeiffer syndrome, and others) (1). In addition to the factors that anatomically increase the respiratory disorders during sleep, some properties of the syndrome increase the morbidity of patients. In Prader–Willi syndrome, in addition to facial dysmorphism, morbid obesity and increased secretions are seen in patients with cerebral palsy, and hypotonia increases the risk of respiratory disorders developing during sleep (2).

In children with Down syndrome, in addition to anatomical factors such as midfacial hypoplasia, micrognathia, narrow nasopharynx, small oral cavity, relative tonsil and adenoid hyperplasia, and laryngotracheal anomalies, increased secretions, hypotonia in the palatal–lingual–pharyngeal muscles, presence of obesity, and presence of other systemic diseases (such as cardiovascular problems) increase obstructive sleep apnea and/or other risks of respiratory disorders developing during sleep (3).

In addition to obstructive sleep apnea and other respiratory disorders, these children have an increased incidence of chronic rhinosinusitis. Further, the recurrent episodes of tonsillitis or adenoiditis affect syndromic children just as they affect other children (3). Respiratory disorders, frequent upper respiratory tract infection, and increased morbidity also occur in patients who have had hepatic or renal transplantation and in patients with diabetes due to metabolic disorders in the presence of decreased immunity and systemic

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problems, in addition to the craniofacial abnormalities. Because the presence of respiratory disorders during sleep in syndromic children and frequent infections in these children will increase the symptoms associated with the cardiorespiratory system and neurodevelopmentally influence these children to a greater extent, the following treatment modalities are recommended to be implemented immediately in syndromic patients with respiratory disorders during sleep (4). These treatment modalities include adenotonsillectomy, midface advancement procedures, hyoid suspension, tracheotomy, surgeries to expand the nasopharyngeal cavity, and the applications of continuous positive airway pressure (CPAP) (1, 4). Surgical interventions to be performed in syndromic patients have increased risks in terms of surgical techniques and anesthesia in both perioperative and postoperative periods. However, surgery can be performed by taking some measures for surgery and anesthesia.

Here we have presented our experiences of tonsillectomy and/or adenoidectomy in syndromic patients who applied to our clinic.

Methods

The study has been approved by the Research Board of Medical and Health Sciences of the Başkent University with the project number KA15/356 and the act 94603339-604.01.02/12897. In this study, we examined the syndromic patients in whom tonsillectomy and/or adenoidectomy was performed by the same surgeon in the otolaryngology clinic of the Başkent University in terms of perioperative and postoperative complications retrospectively between the years 2001 and 2011. The ENT examination notes of the patients having complaints of snoring, nasal congestion, frequent episodes of tonsillitis, and frequent upper respiratory tract infection, and the findings detected in the examination were analyzed. The size of adenoid hypertrophy was determined as the percentage of closure of the choanal apertures by using flexible fiberoptic endoscopy in all patients. Tonsillar hypertrophy was classified according to the rate of tonsil to oropharynx from 0 to +4. According to this: (0) in tonsillar fossa; (+1) 25% of the tonsil; (+2) 25-50% of the tonsil; (+3) 50-75% of the tonsil; (+4) more than 75% in the oropharynx.

Table 1. The characteristics of the patients

Age	Gender	Complaint			Tonsil size	Adenoid size %	Applied surgery	Primary disease
		Snoring	frequent tonsillitis	Others				
3.5	F	+	+	-	+3	90	T+A+VTA*	Down Syndrome
5.5	F	+	+	swallowing difficulty	+4	90	T+A+right VTA	Down Syndrome+Congenital Hypothyroidism
4	F	+	+	-	+3	95	T+A+VTA	Down Syndrome+Hypothyroidism Nephrolithiasis+Repaired VSD+PDA **
2.5	M	+	+	swallowing difficulty + vomiting	+3	90	T+A+VTA	Cerebral palsy+Epilepsy
6	M	+	–	-	0	95	A	Coarctation of the aorta+Hypertension
4.5	M	+	–	hearing loss	0	90	T+A+VTA	nephrotic Syndrome
5	M	+	+	-	+3	90	T+A	Craniosynostosis+Asthma
5	F	+	+	-	+3	90	T+A	Liver Transplantation (Congenital Biliary Atresia)
5.5	M	+	+	-	+3	90	T+A+VTA	Liver Transplantation (tyrosinemia type 1)
5	M	–	–	nasal congestion and discharge	0	95	A	Liver Transplantation 8-Methylcrotonyl CoA Carboxylase Deficiency
7	M	+	+	-	+4	90	T+A+VTA	Renal Transplantation (Chronic Renal Failure)
11	F	+	+	-	+4	90	T+A	Type 1 Diabetes Mellitus
6	M	+	+	-	+3	90	T+A	Type 1 Diabetes Mellitus
7	M	+	+	-	+3	90	T+A	Mediterranean Anemia Carrier

*A: adenoidectomy; T: tonsillectomy; VTA: ventilation tube application

**VSD: ventricular septal defect; PDA: patent ductus arteriosus

Respiratory disorders during sleep were evaluated with a detailed questioning of the family in all patients. The cases of sleeping with open mouth, snoring, and breathing pauses were evaluated. Surgical decision was made in witnessed apneas or in the records taken with mobile phones, widely used in recent years, or cameras by the families during sleep.

Results

The complaints, physical examination findings, surgeries performed, and primary diagnoses of 5 female and 9 male patients aged 2.5 to 11 years are shown in Table 1. While there was a complaint of snoring in all patients, except for one, eight patients also had a history of frequent tonsillitis. The tonsil size was +3/+4 in the examination of patients who underwent tonsillectomy along with adenoidectomy. All patients underwent adenoidectomy and the adenoid size of the patients was between 90% and 95% according to the percentage of closure of the choanal apertures. While tonsillectomy, adenoidectomy, and a ventilation tube insertion were applied in seven patients, tonsillectomy and adenoidectomy were applied in five patients and only adenoidectomy surgery was performed in two patients.

All patients were hospitalized and followed in the postoperative period. Two patients diagnosed with type 1 diabetes mellitus were hospitalized for 3 days and the other patients were hospitalized for 1 day and followed postoperatively. No complications in anesthesia and/or surgery developed in any of the patients. Similarly, in the postoperative period, none of the patients received intensive care.

Discussion

In this retrospective study consisting of 14 syndromic cases, we observed that the complaints of snoring and frequent infection in all the patients were at the forefront. No complications developed in any of the patients in the postoperative period; however, we believe that all patients should be hospitalized and monitored because of potential complications that could occur. Although adenoidectomy and tonsillectomy surgeries are at the forefront, particularly in syndromic children with respiratory disorders during sleep, it should be kept in mind that the surgeries performed in these children may be inadequate and their respiratory disorders may continue during postoperative sleep.

The frequency of obstructive sleep apnea in children is between 1% and 6% and is mostly seen between 2 to 8 years of age (5). Obstructive sleep apnea and/or respiratory disorders during sleep are reported in higher rates in syndromic patients (1, 6). Children with syndromic craniosynostosis are reported to be more prone to obstructive sleep apnea, and these rates are between 40% and 85% (7). Although there is no snoring in these children, it is known that obstructive sleep apnea may be observed (7). Early intervention is recommended in syndromic children because of the increased risk of morbidity and mortality. However, it is not certain when respiratory problems will occur in syndromic children. Sometimes, complaints may begin during the infantile period (1). Treatment modalities vary according to the age of complaint onset. Polysomnography is considered to be the gold standard in the diagnosis of obstructive sleep apnea in syndromic patients. We did not perform polysomnography in patients with snoring complaints in the preoperative and postoperative periods. Therefore, it was not possible for us to objectively determine how much our patients benefited in the postoperative period.

Adenotonsillectomy is still performed as a first step in the treatment of sleep-disordered breathing in children without any disease other than obstructive sleep apnea. Because adenoids and tonsils reach the maximum size between the ages of 3 to 6, excision gives the best result in this age group. In the craniofacial dysostosis syndrome, changes in the mid-facial bone structure, laryngeal and tracheobronchial anomalies can cause airway obstruction during infancy. Therefore, tracheostomy, nasopharyngeal airway, or CPAP can often be the first step of treatment in these patients with complaints that began during infancy (4). Although the surgical correction of mid-facial deformities is regarded as a definitive treatment, it has been observed that the best results are obtained after facial growth is complete (8). However, even though tonsil and adenoid hypertrophy is not excessive in older ages, it was stated that these syndromic patients could benefit from adenotonsillectomy (9). In another study in which adenotonsillar surgery was evaluated with polysomnography in children with syndromic craniosynnostosis diagnosis, it was seen that this surgery did not significantly reduce obstructive sleep apnea (6). Because the children with syndromic craniosynostosis may have mid-facial hypoplasia, narrow oropharyngeal area, pathologies related to upper respiratory tract, and problems with the central nervous system; care in terms of the use of instruments during surgery, especially in the preoperative period and in terms of the possible complications related to anesthesia in the preoperative and/or postoperative period, must be taken (6).

It was seen that the children with Prader–Willi syndrome that have hypotonia and obesity along with facial anatomical abnormalities benefited from adenotonsillectomy in the presence of adenotonsillar hypertrophy; but postoperative complications increased in patients especially under 3 years of age (10). As in our patient, improvement can be achieved especially in swallowing in the presence of adenotonsillar hypertrophy through adenotonsillectomy in children with a disease such as cerebral palsy, which has an abnormal motor tone.

Down syndrome is the most common chromosomal disorder (1:660–1:800) and the incidence of obstructive sleep apnea is reported to be 30-60% in these patients (11). Adenoidectomy and tonsillectomy are often performed in the treatment of ob-

structive sleep apnea and/or respiratory disorders during sleep in patients with Down syndrome, except for recurrent tonsillitis and adenoiditis; however, the surgery may fail because of various anatomical and physiological abnormalities. Persistent or recurrent obstructive sleep problems were reported after adenotonsillectomy surgery in these children at a rate of 30-50% (8). Adenotonsillar surgery is known to carry the risk of increased complications in this group of patients. Abnormalities in anatomical structures, increased secretions, hypotonia, atlantoaxial instability, increased pulmonary vascular resistance, abnormal control of the central nervous system on breathing, increases in upper and lower respiratory infection incidence, laryngotracheal anomalies, and increased risk of subglottic stenosis make the intraoperative intubation and surgery difficult as well as increase the risk of postoperative respiratory complications (3). Because the laryngeal and cricoid cartilage tend to be narrower in patients with Down syndrome, the risk of subglottic stenosis development is known to be high. Again, since they carry the risk of atlantoaxial subluxation as well, the extension of the neck is important for neurological complications that may develop (12). In a study performed, intraoperative anesthesia complication was reported as 8% (especially respiratory complications) (3). The proportion of patients who needed intensive care and postoperative airway control was reported to be 25% in the same study. In another study, major respiratory complications in syndromic patients were reported to be 27% in general (13). Delay in the initiation of oral fluid intake in the postoperative period was another issue that was reported (3). We did not encounter postoperative respiratory complications in our syndromic patients after the adenotonsillectomy. Because retardation may occur in mental development of the children with Down syndrome, hearing becomes even more important in these patients. Therefore, we think that a more liberal attitude is required in inserting the ventilation tube into the eardrum.

Another group of patients in our case series consisted of children with solid organ transplantation. Many observational studies indicated that adenotonsillar hypertrophy was a problem after solid organ transplantation (14, 15). However, because it is a problem that was frequently encountered in the normal population, it has not yet been clarified whether or not solid organ transplantation alone is a risk factor in adenotonsillar hypertrophy. On the other hand, as this group of patients was immunosuppressive, they were exposed to more frequent infections. Frequent infection, especially Epstein-Barr virus (EBV) infection, is thought to be the cause of hyperplasia (16). Another important disease that can occur after organ transplantation is posttransplant lymphoproliferative disorder (PTLD). Especially after renal transplantation, the incidence is reported to be 1.2-10% and mortality may be 10% despite treatment (15, 17). Hyperplasia of the nasopharyngeal lymphoid tissue may be a PTLD precursor as in our case (15). However, the growth of all lymphoid tissues located in Waldeyer's ring could be seen (17). Our patient was a five-year-old child who had undergone a liver transplantation after cryptogenic cirrhosis due to 3-methylcrotonyl CoA carboxylase deficiency. In our clinic, when consulted due to nasal congestion, difficulty breathing through the nose, and an inflamed nasal discharge, there was a mass completely filling the nasopharynx, extending to the nasal cavity, having a hard consistency and bleeding when touched. Curettage adenoidectomy was applied in the patient and the pathology result was B-cell lymphoma. The result of the nasopharynx biopsy demanded control purposes after the chemotherapy was consistent with epithelial hyperplasia. PTLD is often found associated with immunosuppressive drug use, pre-transplant seronegative EBV status, transplantation in childhood, and the use of anti-thymoglobulin and anti-CD3 antibodies (18, 19). Surgery was also recommended in the case of the PTLD suspicion (20). B-cell lymphoma developed in one of our patients after liver transplantation and the diagnosis was made through adenoidectomy. In patients who underwent renal transplantation, tonsillectomy was recommended after transplantation in order to suppress recurrent IgA nephropathy. Tonsillectomy was shown to reduce protein excretion in urine in these patients (21).

It is also known that patients are more vulnerable to infection and their blood glucose regulation is impaired due to infection from other metabolic diseases such as diabetes mellitus. Disorders of the immune system and disorders in leukocyte functions, especially in the case of acidosis, are known in patients diagnosed with Type 1 diabetes mellitus (22). This situation can also increase the incidence of infection in this patient group. It is recommended that patients be carefully monitored postoperatively in this adenotonsillectomy group due to frequent incidence of tonsillitis or sleep-disordered breathing. Although nausea, vomiting, and oral intake disorders are the most common problems in this group of patients, especially after tonsillectomy, because the blood glucose regulation may be significantly impaired in these patients, they should be hospitalized and followed until oral intake is provided. There were two patients with Type 1 diabetes mellitus in our case series. Frequent episodes of tonsillitis experienced by the patients disrupted blood glucose regulation, caused hyperglycemic and hypoglycemic episodes, and their insulin needs increased. The need for insulin significantly decreased in patients who were hospitalized for three days until adequate food intake was ensured after surgery. Information was given that diabetes regulation was started to be provided much more easily after the surgery.

Conclusion

It should be noted that respiratory problems in syndromic patients may arise from many different anatomical regions, and surgery should be performed in these patients taking into account all the areas. Keeping in mind that these patients had upper and lower respiratory tract infections more frequently and that the incidence of respiratory disorders during sleep was higher, adenotonsillectomy should be considered as the first-step therapy in the presence of adenotonsillar hypertrophy. However, it should be kept in mind that there is a chance of failure in adenotonsillectomy because of craniofacial abnormalities and problems in

the other systems in these patients and there might be a need for other additional treatment modalities. In addition, considering the increased risk of complications, patients should be hospitalized and followed in the postoperative period.

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