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Children With Facial Differences: The Problem of Tonsils and Adenoids

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Introduction

Reduced dimensions in the airways of children and fast development of the face can lead to irreversible problems if there is prolonged compromise of breathing patterns. In addition to early socialization, maturation of the immune system can in that period favor recurrent infections in children. These problems become even more complex in children with facial differences. [1,2,3]. The specialist in ENT who works with children with special needs, one of the branches of pediatric ENT, and the team managing syndromic children or children with clefts of the lip and palate, are entangled in a delicate universe of difficult solutions. Thus, understanding the context of these patients is as important as understanding the pathophysiology of their diseases.

Anatomy and physiology applied to tonsils and adenoids

The palatine tonsils and adenoids are under constant immunologic activity, especially during the first years of a child's life. This process can lead to significant reaction and hypertrophy of lymphoid tissue in tonsils and adenoids. The period with greatest activity and physiological enlargement of these organs is between three and ten years of age. Function of the adenoids decreases slowly and progressively during puberty. Similarly, the palatine tonsils undergo involution, but this process happens later, between 15 and 30 years of age. As the adenoid goes through its growth cycle, the nasopharynx suffers changes in shape and volume as a result of growth of the skull and face.

In a child, the nasopharynx has a smaller volume and is flatter in shape; as the child grows it becomes larger and more prominent. In some children with syndromes or with craniofacial malformations the nasopharynx can remain thinner and smaller even as the child grows, and the ratio between volume of adenotonsillar tissue and of the pharynx can have some characteristics that can help or hinder breathing and velopharyngeal functions [4-5]. In addition to obstructions, infectious processes go beyond local conditions and can compromise the physiology of the nose, paranasal sinuses, middle ear and auditory tube complex, and even the esophageal-pharyngeal-laryngeal tract and lower airways. Thus, to define the best approach for a child with a cleft and adenotonsillar problems, physicians must know anatomy, physiology, immunology, pathology and the pattern of growth of adenoids, tonsils, and face.

Main clinical manifestations related to tonsils and adenoids in children with craniofacial malformations

Children with cleft palate or any other facial malformation leading to adenotonsillar hyperplasia have clinical manifestations associated with changes in breathing patterns and in articulation and the organs of speech. Other nasal problems such as allergic rhinitis can also mimic and intensify these manifestations that become more pronounced during the growing of the face, characterizing the **Mouth-Breathing Syndrome of Children with Special Needs**. **Figure 1** shows the impact of adenotonsillar hyperplasia on quality of life in various stages of childhood and adolescence. The manifestations were grouped on the basis of their most frequent onset, and those described in a certain phase that can continue in further years.

Figure 1. Evolution of Clinical Manifestations of Mouth-Breathing Syndrome in Children with Special Needs associated with Adenotonsillar Hyperplasia

0 - 2 years old Noisy breathing and panting, night snoring, obstructive sleep apnea, agitated sleep, difficulty in nursing and breathing, producing snore-like sounds while eating, frequent rhinorrhea, deficit in weight gain. Overprotection or neglect are more frequent in those with syndromes with facial malformations.
2 - 4 years old Lips ajar, drooling, eating with the mouth open, narrowing of the nasal fossae, alterations in teeth, problems with speech, "hot potato" voice, hoarse voice, nocturnal enuresis, delay in growth. They start having problems with self-esteem and behavior.
4 - 6 years old More significant aesthetic alterations of the face, elongated and unexpressive face, postural alterations (anterior projection of head and shoulders), waking up during the night to drink water, headache when waking up, irritability, sleepiness during the day, attention deficit shown in pre-school, poor appetite, lack of enthusiasm for sports, more intense problems with self-esteem.
7 years old and adolescent Problems with writing and performance at school, dry mouth, hyperplasia of the gums, gingivitis, halitosis, dry lips, poor performance in sports, obesity, sleepiness during the day. Facial deformities, alterations in the nasal pyramid especially, become more visible. Problems with speech, especially due to a hypernasal voice, impair socialization. Adolescents complain they have bad breath and can't kiss properly because of a blocked nose.

Airway obstruction

In most cases of respiratory obstruction, adenotonsillar lymphoid tissue occupies a disproportionate amount of space in the upper airways. This obstruction has a dynamic component, with the exacerbation of symptoms when the child is leaning backwards or lying down, in malformations of the jaw (Pierre Robin Sequence and Treacher Collins Syndrome), in macroglossia (Down Syndrome and Beckwith-Wiedmann Syndrome), when there is reduced neuromuscular tonus during sleep, in cerebral palsy, and when the child is using drugs that interfere

with the sleep-wake cycle. Obese children are also at greater risk of having respiratory obstruction during sleep due to collapse of more voluminous soft tissue in the airways. This condition can also be exacerbated by the anatomical narrowing of nasal fossae and pharynx, as observed in some syndromes with craniofacial alterations (Pierre Robin Sequence, Down Syndrome and Treacher Collins Syndrome) [3,6].

Sleep-related breathing disorders – obstructive sleep apnea syndrome

Obstructions of the airways due to adenotonsillar hyperplasia, more visible during sleep, are the primary cause of Sleep-Related Breathing Disorder (SRBD) in normal children. In those with craniofacial malformations, frequently found characteristics such as shorter anterior base of the skull, mandibular and maxillary hypoplasia, and macroglossia reduce the diameter of the airways. Thus, adenotonsillar hyperplasia in children with special needs may be considered a factor that exacerbates obstructions related to the characteristics described previously. In its milder form, SRDB is called Upper Airway Resistance Syndrome (UARS). Children with more significant degrees of obstruction might have the Obstructive Sleep Apnea Hypopnea Syndrome (OSAHS). The SRDB condition is originated primarily during the REM stage of sleep, when children are less observed by their parents. In many cases of OSAS and OSAHS, parents may misinterpret symptoms, underestimating them as simple snoring without obstruction [7,8].

The dynamics of the airways during sleep cannot be determined by a static examination in the consultation room, especially in younger children. Assessment of the nasopharynx, adenoids, and tonsils using fiber optics is indicated to determine choanal and oropharyngeal obstructions, but it will not show changes in the dynamics of the pharynx and tongue during sleep. Similarly, radiographic evaluation of adenoid tissue does not show these alterations, and it does not have adequate sensitivity for judicious assessment of the degree of obstruction.

There is still no consensus on criteria for diagnosing SAOS in children with facial differences, and there is no research correlating facial malformations and polysomnography abnormalities with unfavorable outcomes in SRDB. Polysomnography (PSG) remains the gold standard for objective correlation of abnormalities of ventilation associated with alterations of breathing during sleep. However, the cost and difficulty of conducting PSG in children means that it is an inconvenient method for assessment in pediatrics. It is important to stress that the criteria for diagnosing sleep-disturbances using polysomnography are not the same for children and adults. Other techniques for assessment include audio recording, video recording, and PSG at home. Favorable results have been observed using these methods, but they require further studies. Simplified PSG (overnight oximetry and PSG while napping) has high positive predictive value and low negative predictive value, suggesting that patients with negative results might still require additional tests [8-9].

Children with syndromes who are more severely affected can develop cor pulmonale, right ventricular hypertrophy, alveolar hypoventilation, pulmonary hypertension, pulmonary edema, poor weight gain, and increased risk of

permanent neurological damage and death [10,11]. In clinical practice, when a history of severe obstruction is consistent with clinical findings, the patient can likely benefit from adenotonsillar surgery. Children with facial differences frequently have obstructive manifestations already at birth, so parents may underestimate its meaning and severity. When the history is not consistent with the physical examination, an additional evaluation including some combination of audio and video recording of the child sleeping at night and endoscopy of the airways can help to establish cause and degree of symptoms. Pre-operative PSG can be saved for assessing children with high surgical risk including those with complex malformations of the airways, cardiopulmonary problems, obesity, neurological deficit, a history inconsistent with the physical examination, and those whose parents or doctors do not rely on the results from breathing obstructions recorded when the child was sleeping.

A PSG test is also indicated when sleep-apnea continues after surgery (residual SAOS). Children with facial differences who have an inadequate response to surgery for SAOS might benefit from continuous positive airway pressure (CPAP), or in some cases from using continuous oxygen during sleep. Some of these children may require tracheotomy.

In some syndromes with SAOS (e.g. Crouzon Syndrome, Robin Sequence) associated problems of upper and lower airways can be observed [6].

Mouth-breathing syndrome in children with special needs

A 3-year-old child has almost 90% of its skull developed. The newborn has a 1:8 face:skull ratio and the adult, 1:2. This means that during childhood the face will grow much more than the skull, and the speed of facial growth is greater in the first four years of life. In the natural growing process of muscles, ligaments, and mandible, there is a tendency to grow downwards and forwards. When the frontal complex grows it pushes the nasomaxillary process forward. That is why the face grows forwards and downwards [1,2].

It has been postulated that chronic nasal obstructions associated with adenotonsillar hyperplasia can predispose some children to abnormal facial growth. The theory is that in these children caudal growth of the mandible added to retro-positioning of the tongue may compensate for the absence of airflow through the nose by creating a larger oral airway. This adaptation would then produce an elongated face and an increase in the gonion angle. Lack of contact between tongue and palate leads to a high and narrow palate and consequently to a posterior crossbite. Furthermore, the facial growth tends to emphasize the facial deformity present, and does not compensate it [1,2].

Although there clearly is a correlation between chronic nasal obstruction and alterations in facial growth, a cause/effect relationship has not been established in humans. Differences in results of research using animal models and that on humans arise from the use of inadequate methods to assess the ratio between nasal and oral breathing, as well as from insufficient follow-up of patients in studies and absence of data correlating the volume of nasal airflow and resistance to this flow, according to age, gender, or weight. Data on children with facial differences are even more incomplete [12].

According to data in published reports, we can infer that the Mouth-Breathing Syndrome (MBS) is characterized by alterations in speech and in organs of articulation due to breathing predominantly through the mouth during childhood, usually associated with alterations in the face, positioning of teeth, and bodily posture. It can also be accompanied by cardiopulmonary, endocrine, and nutritional alterations, sleep and behavioral disorders, and poor performance at school. It is related to genetic factors, to non-nutritional sucking habits (pacifiers, bottles, thumbs, and biting of nails) and to nasal obstructions of different degrees and duration. In children with facial differences, frequently those with cleft lip and cleft palate, MBS can lead to even greater problems [12].

Some data suggest that small abnormalities in speech and the organs of articulation and in dentofacial growth in patients with adenotonsillar hyperplasia can be reversible if a nasal breathing pattern is restored. However, late identification and intervention favor the development of irreversible changes in the profile of the face and in the quality of life of children with MBS. An interdisciplinary approach is preferable for controlling the consequences of mouth-breathing. Early diagnosis and intervention in children with special needs who have MBS seems to be even more important. The pediatrician plays an important role in identifying and in conducting the clinical follow up of these children. The specialist in ENT makes the etiologic diagnosis and is concerned with indications for clinical or surgical treatment of the obstruction. The speech therapist works on restoring soft tissues, the orthodontist the dental arch, and the physiotherapist postural changes. A follow up with a nutritionist might also be necessary [12-14]. Other professionals can also be consulted: an allergist, pulmonologist, endocrinologist, and cardiologist.

Changes in voice, speech and swallowing

During the period of growth of the adenoids, velopharyngeal closure can be facilitated by the lymphoid cushion, but breathing can be compromised and can be followed by hyponasal phonation. Adenoid tissue can act as a cushion over the posterior wall of the pharynx, influencing functioning of the velum, limiting muscular effort in swallowing and phonation, and can allow good occlusion in cases of cleft palate. When lymphoid tissue involutes, nasal ventilation may be adequate, but there may be some compromise in phonation resulting in a hypernasal voice and reflux of food through the nose.

Velopharyngeal closure in children with large adenoids occurs by a different mechanism from that in children without adenoids. The hypertrophied adenoids can ease the action of an insufficient palate or can hide other deficiencies that will appear progressively with physiological atrophy of the adenoids or appear dramatically after an adenoidectomy. The changes observed in velopharyngeal closure after this surgery require a much stronger muscular effort.

Hypertrophied tonsils can also interfere with the function of this malformed palate. Those accompanied by a more developed superior pole, which occupies a larger volume of the supra-tonsillar fossa, cause greater limitation of movement of the soft palate, dislocating it upwards and backwards. Surgical removal of these tonsils can lead to fibrosis inside the velum, decreasing its elasticity and compromising even more the child's phonation and velopharyngeal closure.

In some cases, mild or moderate hyperplasia of lymphoid tissue can favor the phonating function of the soft palate and stop oronasal reflux. So it may be complex to determine the role lymphoid tissue plays in symptoms of children with velopharyngeal incompetence or insufficiency and adenotonsillar hyperplasia.

Tonsillar and/or adenoidal hyperplasia can also cause dysphagia by interfering with the pharyngeal phase of swallowing. For these children, it is harder to swallow solid foods than liquids. They usually chew with their mouth open. Severe tonsillar hyperplasia in children with a normal palate rarely causes problems with velopharyngeal closure, so nasal regurgitation is not frequent in these cases. Children with clefts and with dysphagia associated with poor weight gain can benefit from adenotonsillectomy. Changes to an upper percentile of weight can be observed in children after this operation. In these cases, before surgery it is essential to define the role of adenoids and tonsils on velopharyngeal function.

Endoscopic assessment of velopharyngeal function is imperative for good surgical programming. The goal of visualizing the nasal face of the palatal velum is to measure and locate areas where air may escape during phonation, to identify the height and pattern of palatal closing, the participation of adenoids, and to rule out the presence of a submucosal cleft. Evaluation made together with the speech therapist can supply more complete information on velopharyngeal function, especially about characteristics such as closure on phonation and swallowing. In nursing babies, this assessment of velopharyngeal closure is difficult to do, and even some young children do not collaborate adequately for the study of all functions of the palate because the endoscope can be quite uncomfortable [12-14]. In some cases, videofluoroscopy can supply more precise information.

In addition to imaging, questionnaires for the assessment of quality of life are an important tool in evaluating the influence of a cleft palate on the child's and the family's daily lives. Children with velopharyngeal insufficiency have problems with functional evaluation and socialization, and parents report further emotional problems [15].

Surgery on the tonsils and adenoids in children with cleft palate

Adenotonsillectomies are among the most commonly performed surgeries worldwide. Many studies have defined populations that are good candidates for this procedure, to avoid loss of immunologically competent tissue and unnecessary surgical risks. When adequately indicated in cases of obstruction, this procedure undoubtedly improves the quality of life for children with facial malformations and in some cases, such as SAOS, it can save lives. Recurrent otitis and sinusitis can also benefit from adenoidectomy. Recurrent tonsillitis is also an indication for surgery.

Surgery on the tonsils and adenoids requires careful evaluation of children with facial differences or with other malformations. Planning and performing this kind of surgery requires special care if there are risk factors: visible or hidden submucosal cleft palate, uvula bifida, palatal hypotonicity or paralysis, craniofacial malformations, cervical vascular anomalies (Velocardiofacial syndrome), anomalies in cervical vertebrae with instability of the neck (Down Syndrome), nasal reflux of fluids, and hypernasal voice [3-6]. The major and minor risks of this surgery

are intensified in syndromic children and in children with neurological deficit or with facial malformations. The most common complications are immediate hemorrhage, post-operative hemorrhage, dehydration, post-operative edema of the airways, scarring with stenosis or adhesions in the upper airways, increase in velopharyngeal dysfunction, and complications of anesthesia. Complications are less frequent and are minor when only the adenoids are removed.

Children remain in the hospital for 12 to 24 hours, and most of them have a smooth recovery [3-6]. Some syndromic children with mandibular malformations or muscular hypotonia, and those with achondroplasia or Down syndrome, undergoing surgery on the adenoids and tonsils have increased risk of postoperative respiratory distress and should be admitted to the intensive care unit after the surgical procedure [3-6,10,11].

No increase in diseases of the immune system has been observed in children who undergo surgeries on the adenoids or tonsils [4].

The child's recovery is fast, although there are individual variations. Children may have some trouble feeding in the first days after tonsillectomy because of the pain. Good analgesic treatment and the feeding of fluids or soft foods may help. Juice, tea, and ice cream are usually well accepted.

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