REVIEW ARTICLE

Hearing impairment in children with cleft lip and cleft palate

Alterações auditivas em crianças portadoras de fissuras labiopalatinas

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ABSTRACT

Cleft lip and cleft palate are congenital malformations that are characterized by cosmetic facial deformity, speech disorders, and conductive hearing loss caused by chronic Eustachian tube dysfunction. There are treatment protocols for the repair of lip and palate that are usually performed between three and 18 months of age, respectively. Palate repair restores palate functions and anatomy, and the floor of the nasal cavity-promoting proper functioning of the Eustachian tube. The correct insertion of tensor muscles and soft palate lift in the hard palate can promote more efficient opening of the Eustachian tube during swallowing, keeping aeration in the middle ear, minimizing the risk of developing otitis media with effusion. The protocols are well-defined about the timing for palatoplasty. However, there is no consensus on the treatment of Eustachian tube dysfunction. Several studies have emerged in order to guide treatment and determine the best time to resolve the disease, choosing to carry out tympanostomy with the placement of ventilation tubes. Early tympanostomy, at four months of age, is indicated by some researchers; however, others are conservative and await the onset of symptoms to intervene surgically. In this study, the current treatment of otitis media with effusion, studies with computer models, the natural history of hearing loss in patients with non-operated cleft lip and cleft palate, and results of ventilation tubes placement on auditory development and speech of this population are evaluated.

Key words: Cleft Palate; Eustachian Tube; Middle Aer Ventilation; Otitis; Hearing.

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RESUMO

As fissuras labiopalatinas são malformações congênitas que têm como características a deformidade estética facial, alterações de fala e disacusia condutiva ocasionada pela disfunção crônica da tuba auditiva. Existem protocolos de tratamento para a reparação do lábio e do palato, que habitualmente são realizados dos três e aos 18 meses de idade, respectivamente. A reparação do palato tem como funções restabelecer a anatomia do palato e assoalho da cavidade nasal e promover o funcionamento adequado da tuba auditiva. A correta inserção dos músculos tensor e elevador do véu palatino no palato duro pode promover abertura mais eficiente da tuba auditiva, durante a deglutição, mantendo a aeração da orelha média, minimizando o risco do desenvolvimento da otite média com efusão. Os protocolos definem bem o que fazer em relação ao momento da palatoplastia, entretanto, não existe consenso sobre o tratamento da disfunção da tuba auditiva. Vários estudos têm surgido com o intuito de orientar o tratamento e definir o melhor momento para a resolução dessa doença, tendo como escolha a realização de timpanotomia com colocação de tubos de ventilação. A timpanotomia precoce, aos quatro meses de idade, é indicada por alguns pesquisadores, entretanto, outros são conservadores e aguardam o surgimento dos sintomas para intervir cirurgicamente. Neste trabalho, verifica-se o que

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existe de atual no tratamento das otites médias com efusão, os estudos com modelos computacionais, a história natural das alterações auditivas em pacientes com fissuras labiopalatinas não operadas e o resultado das introduções de tubos de ventilação no desenvolvimento auditivo e de fala dessa população.

Palavras-chave: Fissura Palatina; Tuba Auditiva; Ventilação da Orelha Média; Otite; Audição.

INTRODUCTION

Cleft lip and cleft palate (CLP) are congenital malformations in which the two side halves of the palate do not fuse during the embryonic development, between the fourth and 12th weeks of pregnancy. These defects are common among those affecting the human face, with an average prevalence of one and two individuals per 1,000 births.[1] In Brazil, this rate oscillates around 1:650[2], which is close to the epidemiological data from European and American white populations ranging between 1: 500 and 1: 768 births, respectively.[3]

Children with FLP have a high prevalence of otitis media with effusion,[4] a disease that determines conductive hearing loss, from mild to moderate, often found in children of preschool and school age. This age is critical in the acquisition of spoken and written language, relationships, and social interactions.[4]

The specialized literature shows a wide variety of studies related to the treatment of otitis media with effusion (OME) in children with FLP. These studies are carried out to develop treatment strategies that can minimize auditory effects and structural sequelae in the tympanic membrane and ossicular chain. Some authors recommend medical and surgical treatment with the introduction of ventilation tubes early, before the onset of signs or symptoms of OME, others adopt a conservative approach, waiting for dysfunctional signs in the Eustachian tube to indicate treatment.

It must also be considered that, as a result of communication and learning difficulties, the impact of this malformation on the quality of life of children and their families is significant. This study aims to review what has been accomplished in highly prestigious centers for the treatment of craniofacial anomalies.

MULTIDISCIPLINARY TEAM

The World Health Organization recommends that services assisting patients with FLP must have a multidisciplinary team composed of a psychologist, nurse, surgeon, orthodontist, speech therapist, otorhinolaryngologist, geneticist, social work (displacement costings), and dentist; and that the skill and experience in their approach requires palate surgeons, orthodontists, and speech therapists to care for at least 40-50 new cases with this malformation per year.[5]

PATHOPHYSIOLOGY OF OTITIS MEDIA WITH EFFUSION IN PATIENTS WITH CLEFT PALATE

FLP and cleft palate cases result in an abnormal facial appearance and speech defects.[1] Moreover, the presence of a cleft palate also results in abnormal insertion and function of the tensor muscle in the soft palate, which is essential for the physiological maintenance of the Eustachian tube (TA).[6] Due to the lack of middle ear ventilation, sterile liquid accumulates inside leading to the installation of chronic otitis media with effusion, which is responsible for conductive hearing loss, from mild to moderate, and reversible. This group of patients is very vulnerable to hearing health and must be treated early because a perfect auditory pathway is essential for the acquisition of the spoken and written language.[4,6]

DIAGNOSIS AND TREATMENT OF OTITIS MEDIA WITH EFFUSION

The early diagnosis and treatment of otitis, especially with the insertion of ventilation tubes, seems to be the main conduct for preventing hearing loss and its functional, social, and psychological consequences.[6]

In 2004, the American Academy of Pediatrics, Family Medical Academy, and the American Academy of Otorhinolaryngology and Neck and Facial Surgery created the Subcommittee on Otitis Media with Effusion (OME), composed of experts in primary care, otorhinolaryngology, infectious diseases, epidemiology, and hearing, speech, language, and nursing care to review the guidelines for the treatment of this condition. This document defines OME as the presence of fluid in the middle ear without signs or symptoms of acute infection. This persistent fluid in the middle ear results in the decreased mobility of the tympanic membrane and acts as a barrier to the conduction of sound. It may occur due to poor TA function or as an inflammatory response following acute otitis media.

About 90% of children have OME before school age, most often between six months and four years of age. Most episodes resolve spontaneously within three months. However, OME is recurrent in 30 to 40% of children; 5 to 10% of episodes take a year or more to clear. Children who are at increased risk of developing OME, including those with FLP, need more readily interventions. The treatment of these children should include hearing tests, evaluation of speech and language (including speech therapy and language concomitant with the treatment of OME), hearing care including sound amplification if required and regardless of OME, and tympanostomy with the insertion of ventilation tubes (TV). After the OME resolution, the child should be resubmitted to hearing tests because the fluid in the middle ear can mask the partial hearing loss and delay its diagnosis.[5]

COMPUTATIONAL MODELS

Sheer et al..[7] used a modeling platform called finite analysis models (MAF), which reproduces the anatomical data of FLP patients and its functional consequences. These models reproduce the complex anatomy/morphology of various elements in the tissue of infants with 3D FLP, and thus, directly simulate the complex physical phenomena (tissue deformation and fluid/air flow) that regulate the function of the Eustachian tube.

Computer models are important to investigate how specific isolated alterations and biomechanical properties influence the TA tube function. Furthermore, the sensitivity of the TA function to alterations in strength in the tensor muscle of the soft palate documented by the model may have important implications for surgical procedures.[7]

NATURAL HISTORY OF HEARING LOSS IN PATIENTS NOT UNDERGOING PALATE SURGICAL RECOVERY

The protocol for FLP treatment in locations where there is research on the subject determines that the palate repair surgery is performed between 12 and 18 months of age. For this reason, it is rare to find people with non-operated palate beyond their first decade of life. Wei Zeng et al.[8] reported that in developing countries such as China, it is common to find people over 20 years old with FLP and non-operated palate, mainly in the southwestern rural areas of that country.

In 2009, these researchers studied the natural history of audiological and tympanometric findings in patients with non-operated FLP. Their goal was to present the audiological and tympanometric profile of 552 individuals, of which 115 were over 10 years old with the non-operated palate, with or without cleft lip. They evaluated 508 ears, of which 54% had hearing loss of more than 15 dB, 23% with type B tympanometric curves, and 11% with type C curves.[8]

PALATOPLASTY AND TA FUNCTION

Alper et al.[9] carried out a study on TA function after palatoplasty in children with FLP based on the application of the forced response test (TRF), which consists of the application of air flow in the middle ear of children with tympanic membrane or with inserted TV to increase pressure up to the point when they are able to maintain passively the Eustachian tube open. Pressure measurements were performed before swallowing and the maximum pressure during swallowing. Test variables were represented by passive TA features (opening pressure, closing pressure, and passive resistance) and by the muscle-assisted tuba active characteristics (TA dilation and constriction, active resistance, and dilation efficiency). The average age of palatoplasty was 14.3 months and the average time between palatoplasty and application of TRF was 4.4 months. The average age at the time of testing was 18.6 months.

The percentage of ears that had increased transtube flow of air during swallowing was 60%. The active resistance and dilation efficiency were similar to those of the normal adult population and older children who underwent tympanostomy for the insertion of ventilation tubes due to OME. However, in studies with post-palatoplasty children who had undergone tympanostomy for the insertion of ventilation tubes in the pre-palatoplasty, the recorded average values were lower.[9]

OME TREATMENT IN CHILDREN WITH FLP

There is a wide variation in protocols used for the treatment of children with FLP, one of them being the routine insertion of TV during palatoplasty or, selectively, in a different time if the symptomatic disease of the middle ear develops.

Phua et al.[10] performed a retrospective study in 2008 with 234 patients with FLP in which children were subjected to palatoplasty from 1990 to 2005. The indication for TV insertion was adopted when there was clinical evidence of OME associated with one the following alterations:

- recurrent episodes of otitis media (more than three episodes in six months);
- audiological evidence of hearing loss greater than 30 dB;
- subjective hearing loss reported by parents (when audiometry had not been performed).

Children with type B tympanogram curve, mild or borderline hearing loss, without acute otitis media, and no subjective hearing loss were treated only with observation.[10]

It was found that most children underwent cheiloplasty before the age of three months, and those with cleft palate underwent reconstructive surgery of the soft palate and/or hard palate between eight and 14 months of age. Out of these 234, more than half underwent at least one tympanostomy with TV insertion. The routine insertion of TV at the time when palatoplasty was performed occurred in 45 (19%) patients; this insertion was not conducted in 189 (80%) patients. TVs were subsequently required in 79 of these 189 patients (41.8%) due to symptomatic disease or middle ear hearing loss. The average was 1.8 procedure in those who routinely received TVs compared to 0.55 of those selected to receive it.[10] Children who routinely received TVs showed worse audiologic and otologic results when undergoing palatoplasty because in a high number of TV inserts led to a high incidence of abnormalities in the tympanic membrane by otoscopy (tympano sclerosis, residual perforation, and consequent hearing loss). The basis of the study recommends that children with FLP only receive TV if clinically indicated by recurrent infections or significant hearing loss.[10]

Flynn et al.[11] compared the prevalence of otitis media with effusion in children with and without FLP. Two groups of children were prospectively followed up from one to five years of age. There were 22 children with unilateral FLP and 21 without a cleft. Cohorts of FLP children born between 1997 and 2002 in western Sweden, and children in the group without clefts born in 2001 in Gothenburg, were evaluated. Children with cleft lip were subjected to closure and subsequent palatoplasty with an average age of 4.3 months (between three and six months) and closing of the hard palate with an average age of 23.28 months (between 11.56 and 37.06 months). Data were collected when they were one-year-old, one and a half years old, and three and five years old. Otomicroscopy, tympanometry, hearing sensitivity (500, 1,000, 2,000, 4,000 Hz), and OME (abnormal tympanometry, in situ TV, otomicroscopy, and abnormal auditory sensitivity) were evaluated.[11] This study demonstrates a significantly higher prevalence of OME in children with FLP than in those without clefts, aged between one and five years. They also presented a higher incidence of tympanostomy with TV insertion than the group without clefts, which indicates increased persistence of OME. In Sweden, tympanostomies with TV placement are generally carried out following up OME for at least three months. When OME was identified, children in both groups showed mild hearing loss. However, those in the FLP group showed higher levels of hearing loss than those without FLP.[11]

FUNCTIONAL SPEECH AND PALATOPLASTY

Merrick et al.[12] in the UK in 2007 assessed the correlation between speech development and functional palatoplasty combined with simultaneous surgical treatment of otitis media with effusion in children born with FLP. A total of 50 patients who had been treated at the center by a single surgeon were examined. Upon palatoplasty, usually between six and nine months of age and when there was evidence of otitis, myringotomy was performed, and TV was inserted. During follow-up, children with recurrent otitis were subjected to other myringotomies with TV insertion. In this study, the incidence of otitis media in children with FLP was 24% (12/50), with no significant difference with the control group, which was 14% (7/50). This result is low compared to previous studies. They attributed this low incidence to both palatoplasty and early TV insertion methods to OME. Functional palatoplasty restores the anatomic continuity of muscles, with the potential to improve the function of the tensor muscle in the soft palate and TA function. The combination of an early insertion of TVs and repeating the insertion as indicated can theoretically reduce the incidence of OME improving hearing in the critical period of speech development.[12]

MASTOID AND TV DEVELOPMENT

In Finland, in a prospective study between 1983 and 1993, Valtonen et al.[13] investigated whether TV placement in children with FLP, with or without cleft lip and before seven months of age, would influence the development of the mastoid during concurrent OME. The patients were examined by experienced otolaryngologists and submitted to microscopy and mastoid x-rays. During this period, 51 children were born with FLP, who were followed-up for six years. The researchers inferred that the FLP group received tympanostomy more often than the control group (a group of children without FLP diagnosed with OME until six months of age who received TV before seven months of age). The results showed that in children with FLP and OME, the early tympanostomy associated with reconstructive surgery of the palate, followed up by a doctor, and repeated TV insertions when necessary seemed to be the ideal treatment. The active treatment strategy decreased potential complications that could occur in tympanostomy with long term tubes and allowed the mastoid development close to normal while maintaining the physiological function of the middle ear in the patient with cleft.[13]

Klockar and Rault[14] at the University of Helsinki recently conducted a study of 97 children with unilateral FLP and evaluated the functioning of TVs placed during the first surgery, at four months of age (while repairing lip and soft palate), and at 12 months of age (during surgery to repair the hard palate). Patients were divided into two groups: with lip and soft palate closure at 3-4 months of age, and closure of the hard palate at 12 months of age; and lip closure at 3-4 months of age and closure of the soft and hard palate at 12 months of age. It was found that the closure of the soft palate at four months of age increases the efficiency of TV insertion and TA effectiveness reducing cases of otorrhea and tube occlusion. Evidence showed that the majority (63%) of children benefited from the early placement of TVs, at four months of age, and that this group was even larger (86%) with the early closure of soft palate.[14]

TREATMENT OF TYMPANIC PERFORATIONS

The treatment of perforations in tympanic membranes in patients with craniofacial anomalies and FLP was evaluated in a study to show the anatomical and functional success of myringoplasty in this group and compare it with the control group. Szabo et al.[13] showed that 98% of children with FLP underwent, at least, one surgery for TV insertion at the age of five. Middle ear repeated diseases and frequent myringotomies for TV insertion increase the risk of perforation of tympanic membranes and, consequently, the need for myringoplasties.

The TA function in children with FLP improves with age, facial skeleton growth, and repair of cleft palate.[15] Smith et al.[16] obtained that the average time to recovery of the Eustachian tube function after palatoplasty was six years, and 79% of patients showed normal TA function after 12 years of age. Hartzel et al.[17] recommend myringoplasty at around 6-7 years of age in children with FLP. However, other researchers suggest postponing surgery until when there is no more Eustachian tube dysfunction signals, after 12 years of age. In some cases, they advise for sub-annular T TV placement to maintain middle ear ventilation.

Ponduri et al.[18] reviewed all studies describing the association between the early TV insertion and its subsequent results in children with FLP. They identified 18 studies, including a randomized controlled trial. The researchers reported that all studies were small and generally of poor quality. There is insufficient strong primary data that provide sufficient evidence base to determine when the routine early TV insertion in patients with FLP has long-term benefits for hearing, speech, language, and psychosocial development.[18]

CONCLUSIONS

The newborn with some orofacial deformity arouses emotions and actions that result in the care from a broad range of health professionals. The well-being of these children involves surgical and non-surgical care, which results in significant costs to public health in addition to the psychological aspects for patients and their families with important social implications. Studies for prevention strategies are emerging. However, the current treatment depends on corrective procedures of the various affected structures in orofacial clefts.[18]

So far there is no consensus on the timing of surgical intervention in patients with FLP in order to avoid hearing damage and sequela in tympanic membranes. The best way to conduct treatment in children with FLP is to use all available knowledge to improve surgical techniques related to palate repair

and tympanostomy for the insertion of TV. Palate repair techniques need to consider the best placement of tensor muscles and to lift the soft palate to allow the most efficient TA functioning, keeping the middle ear cavity aerated. TVs should be inserted taking into account the status of the middle ear and tympanic membranes, being introduced early and/or selected according to the needs.

The feasibility of tube insertion must also be considered because the external auditory canal in young children is very small, which greatly hinders the handling of tympanic membranes. A multidisciplinary treatment is crucial and must involve Dentistry, Speech Therapy, Social work, Nursing, Nutrition, Genetics, Surgery, Otorhinolaryngology, and Psychology. Treatment should be gentle and compassionate towards patients and their families to alleviate suffering, involving all with tranquility so that the child with FLP overcome all stages of his development without traumas.

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