Braz J Otorhinolaryngol. 2017;83(4):464–474

Brazilian Journal of
OTORHINOLARYNGOLOGY
www.bjorl.org

REVIEW ARTICLE

Hearing thresholds at high frequency in patients with cystic fibrosis: a systematic review☆

Debora T.M. Caumo[a,*], **Lúcia B. Geyer**[b], **Adriane R. Teixeira**[c], **Sérgio S.M. Barreto**[d]

[a] *Universidade Federal do Rio Grande do Sul (UFRGS), Faculdade de Medicina (Famed), Saúde da Criança e do Adolescente, Porto Alegre, RS, Brazil*
[b] *Hospital de Clínicas de Porto Alegre (HCPA), Serviço de Otorrinolaringologia, Porto Alegre, RS, Brazil*
[c] *Universidade Federal do Rio Grande do Sul (UFRGS), Instituto de Psicologia, Departamento de Saúde e Comunicação Humana, Porto Alegre, RS, Brazil*
[d] *Universidade Federal do Rio Grande do Sul (UFRGS), Faculdade de Medicina (Famed), Departamento de Medicina Interna, Porto Alegre, RS, Brazil*

Received 16 June 2016; accepted 17 October 2016
Available online 9 November 2016

KEYWORDS
Cystic fibrosis;
Audiometry;
Ototoxicity;
Hearing loss

Abstract
Introduction: High-frequency audiometry may contribute to the early detection of hearing loss caused by ototoxic medications. Many ototoxic drugs are widely used in the treatment of patients with cystic fibrosis. Early detection of hearing loss should allow known harmful drugs to be identified before the damage affects speech frequencies. The damage caused by ototoxicity is irreversible, resulting in important social and psychological consequences. In children, hearing loss, even when restricted to high frequencies, can affect the development of language.
Objective: To investigate the efficacy and effectiveness of hearing monitoring through high-frequency audiometry in pediatric patients with cystic fibrosis.
Methods: Electronic databases PubMed, MedLine, Web of Science and LILACS were searched, from January to November 2015. The selected studies included those in which high-frequency audiometry was performed in patients with cystic fibrosis, undergoing treatment with ototoxic drugs and published in Portuguese, English and Spanish. The GRADE system was chosen for the evaluation of the methodological quality of the articles.
Results: During the search process carried out from January 2015 to November 2015, 512 publications were identified, of which 250 were found in PubMed, 118 in MedLine, 142 in Web of Science and 2 in LILACS. Of these, nine articles were selected.

☆ Please cite this article as: Caumo DT, Geyer LB, Teixeira AR, Barreto SS. Hearing thresholds at high frequency in patients with cystic fibrosis: a systematic review. Braz J Otorhinolaryngol. 2017;83:464–74.

* Corresponding author.
E-mail: deboratmc@hotmail.com (D.T. Caumo).

Peer Review under the responsibility of Associação Brasileira de Otorrinolaringologia e Cirurgia Cervico-Facial.

http://dx.doi.org/10.1016/j.bjorl.2016.10.006

Conclusion: The incidence of hearing loss was identified at high frequencies in cystic fibrosis patients without hearing complaints. It is assumed that high-frequency audiometry can be an early diagnostic method to be recommended for hearing investigation of patients at risk of ototoxicity.

PALAVRAS-CHAVE
Fibrose cística;
Audiometria;
Ototoxicidade;
Perda auditiva

Limiares auditivos em altas frequências em pacientes com fibrose cística: revisão sistemática

Resumo
Introdução: A audiometria de altas frequências pode contribuir para a detecção precoce de alterações auditivas causadas por medicações ototóxicas. No tratamento dos pacientes com fibrose cística, existem muitos fármacos ototóxicos amplamente utilizados. A detecção precoce de alterações auditivas deve permitir que sejam identificadas antes que o dano atinja as frequências da fala. A lesão causada pela ototoxicidade é irreversível, traz importantes consequências sociais e psicológicas. Nas crianças, a perda auditiva, mesmo restrita às altas frequências, pode afetar o desenvolvimento da linguagem.
Objetivo: Investigar a eficácia e a efetividade do monitoramento da audição por meio da audiometria de altas frequências em pacientes pediátricos com fibrose cística.
Método: Foram consultadas as bases de dados eletrônicas PubMed, Medline, *Web of Science* e Lilacs, de janeiro a novembro de 2015. Foram selecionados os estudos em que foi feita audiometria de altas frequências em pacientes com fibrose cística em tratamento com medicamentos ototóxicos e publicados em português, inglês e espanhol. Para a avaliação da qualidade metodológica dos artigos optou-se pela utilização do Sistema Grade.
Resultados: No processo de busca feito de janeiro de 2015 a novembro de 2015 foram encontradas 512 publicações, sendo 250 na PubMed, 118 na Medline, 142 na *Web of Science* e dois na Lilacs. Desses, foram selecionados nove artigos.
Conclusão: Identificou-se a ocorrência de perda auditiva em altas frequências em pacientes com fibrose cística sem queixas auditivas. Admite-se que audiometria em altas frequências possa ser um método de diagnóstico precoce a ser recomendado para investigação auditiva de pacientes em risco de ototoxicidade.

Introduction

Cystic fibrosis (CF), also called mucoviscidosis, is caused by mutations in a gene located on the long arm of chromosome 7 (7q31). This gene encodes the cystic fibrosis transmembrane conductance regulator (CFTR), which functions as a chloride channel, performing and controlling ion transportation through the cell membrane.[1]

Individuals with CF have chloride secretion failure in the respiratory epithelium, which results in excessive sodium absorption, resulting in increased influx of water into cells and therefore elevating mucus viscosity. The mucus becomes approximately 30–60 times thicker than normal. It does not directly affect ciliary beat, but ciliary action becomes ineffective in the clearance of such highly viscous substance, causing stasis, predisposing to ostial obstruction and increased bacterial colonization.[2,3] This can lead to recurrent pulmonary infections, chronic obstructive pulmonary disease, sinusitis, nasal polyps, gastrointestinal malabsorption secondary to pancreatic failure, meconium ileus, rectal prolapse and infertility due to obstruction of the vas deferens.[2]

The incidence of CF varies according to ethnicity, occurring more commonly in Caucasians. Being an autosomal recessive disease, when each parent has the gene for CF, the probability of the birth of a child with the disease is 25%. In accessing ethnic groups, the incidence of CF varies from 1/2000 to 1/5000 live births among Caucasians in Europe, the United States and Canada, 1/15,000 in African-Americans, and 1/40,000 in Finland.[4,5]

In Brazil, the estimated incidence for the southern region is closer to that of the Central European Caucasian population, whereas for the other regions, it decreases to approximately 1/10,000 live births.[4] The prevalence of the mutation in the CFTR gene (ΔF508), in the pediatric population of patients with CF born in Porto Alegre is 60%, with an estimated incidence of 10 cases a year in the city.[6]