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Journal of Otolaryngology-ENT Research

Case Report

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Audiological aspects of "monge deafness"

Volume 6 Issue 5 - 2017

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Received: July 09, 2016 | **Published:** April 11, 2017

Introduction

Hereditary hearing loss of Carthage Monge family is a genetic disorder that affects many families in our country, most of the members reside in the province of Cartago, Taras, Quircot, El Molino and other neighborhoods, however, the author has records of several patients residing in other areas as Pavas or Guápiles. It is time that the national audiological community takes corrective action in the treatment and rehabilitation of patients with this disorder, since technologies now allow better solutions to individuals suffering from this scourge.

History

Are records of hearing loss known as hereditary deafness Monge from about two centuries ago. By oral tradition known to doctors as the Carthaginians Dr. Jesús Guzmán studied this disorder since the early twentieth century. In 1977, researchers at the University of Costa Rica began the study and characterization of this hearing loss, these studies led to the construction of the first tree and connects with a common ancestor named Felix Monge, who was born in Carthage in 1740, apparently wealthy and prolific, stated in his will that he was deaf.

In 1991, it was identified that the Monge family, genetic damage is located on the long arm of chromosome 5. Such damage was appointed by the International Commission of gene nomenclature as DFNA1 (DFN is an abbreviation of deafness word deafness in English and A1, which means prevail autosomal dominant gene). Today there are over 100 individuals carriers of this disorder.

Genetics

Hearing loss of Monge, is a genetic disorder that is inherited from parents to children. It is autosomal dominant. Autonomy means that children of both sexes inherited no difference, i.e. it is not sex-linked. Dominant means that only one copy of the mutated gene altered or hearing loss occurs. Those affected have a copy of the mutated gene and an intact copy. This means that when an affected individual has children with a hearing individual can inherit mutated copy or copy intact. Children who inherit a mutated copy of the deaf parent have hearing loss at some point in life.

When an affected individual has children, there is a 50% chance of inheriting the mutated copy and 50% of that inherits the intact copy. If intact copy inherits the child will not convey listener and hearing impairment. A mutated copy of the gene is sufficient to present the alteration as this hearing loss is dominant. No cases of hearing children hearing loss transmit to their offspring are known.[1] The Monge-syndromic deafness is not because no known associated anomalies. The researchers propose that the mutation affects cytoskeletal repair of the sensory cells of the ear, including actin cilia of the hair cells of the organ of Corti within the cochlea. When these structures damaged, no gates which allow passage of salts (especially potassium) and this in turn causes the neuroelectric currents that carry information to the brain will not occur open.[2]

Audiological studies

Hearing tests conducted over the years from the baseline to allow today enumerate certain characteristics:

a. Sensorineural hearing loss
b. Progressive
c. Hereditary
d. Nonsyndromic
e. Starts in childhood, approximately between 4 and 6years
f. Primarily affects the low frequencies and as it progresses, all frequencies in the audible range
g. The cochlear lesion
h. The post lingual hearing loss is
i. The use of hearing aids accelerates degeneration stereocilia cochlear hair cells.

Hearing tests performed by the author have demonstrated that the use of headphones in one ear, causes the fastest on the side adapted impairment, where that is a cause to contraindicate its use, since not using prosthesis causes a deprivation finally generates aural and auditory processing problem at the time that the patient decides to use the prosthesis makes it very difficult for this disorder. Another myth ruled today is the accelerated evolution with pregnancies, as in monitoring pregnant carrier has not demonstrated an evolution of deafness.[3]

Prosthetic hearing rehabilitation

Note that to alleviate the devastating psychological, sociological and physiological effects of hearing loss hereditara is vital early identification of deafness Monge. This is easy to achieve if they are made from childhood hearing tests to all children of patients with hearing impairment. Unfortunately today there is no program to monitor all families with members carrying damage.[4] The author suggests that in the early stages, when the hearing loss is expressed with bilateral sensor neural fall mild to moderate low frequency, patients should be provided with wireless communication equipment for school use, this will improve the signal to noise ratio within the classroom and will prevent the secondary auditory processing disorder

to poor discrimination in noise and lack of acoustic energy in the perception of speech sounds. This would improve school performance in children carrying early stage.[5]

Once actually affect hearing loss frequencies of 1000 and 2000 Hertz, the adaptive digital, programmable hearing aid with at least two channels in order to handle the frequency disparity is proposed perception.[6] To obviate the aesthetics alleged by the patients with hearing loss, adaptation peritimpánico prosthesis type is suggested, as long as the age and size of the external auditory canal permit.[7] When the patient reaches adulthood and hearing loss progresses to profound hearing loss level, the performance of cochlear implant technology available through social security, which replaces the function of the hair cells in the cochlea (where it has proposed degeneration).[8] Carriers of the Monge hearing loss, are ideal candidates for this procedure because of their status have acquired hearing loss after the acquisition of language. Figure 1 Evolución audiométrica de la hipoacusia de la familia Monge.[9,10]

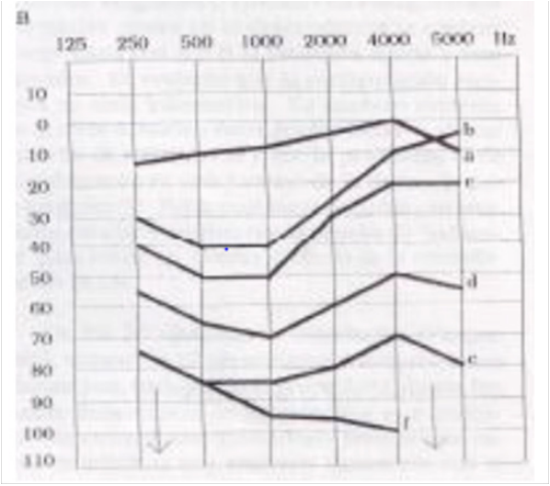

Figure 1 Evolución audiométrica de la hipoacusia de la familia Monge (Según Sánchez).

Conclusion

The implementation of a rehabilitation program for patients with Monge Deafness is essential in order to minimize the adverse effects of this disorder, which are completely preventable by timely identification. Institutions such as the University of Santa Paula, the Costa Rican Social Security and distributors of hearing aids have the responsibility to initiate a program of detection and rehabilitation of hearing deafness due to Monge. Meanwhile audiologists, otolaryngologists and students observe impassively as compatriots uselessly sacrificed their lives fighting a disability that can be rehabilitated, the problem continues to advance and raising its cost to social, economic and quality of life for individuals. The Monge have the right and should have access to technology and surgical solutions that represent auditory and cochlear implant prostheses to reduce the negative effects of this disease.

Acknowledgments

None.

Conflicts of interest

Author declares there are no conflicts of interest.

Funding

None.

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