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Patterns of hearing loss in patients with primary immunodeficiency

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A B S T R A C T

Primary immune deficiency refers to a heterogeneous group of abnormalities in which there are one or more defects in the group of immune system's components. Several mechanisms can be expected for auditory sensory conduction defects associated with immune deficiency. Therefore, the purpose of this study is to investigate the loss of hearing in patients with primary immune defects, using the hearing test (Audio Tympanometry, OAE, ABR) patterns of low sensory neural hearing are classified into a variety of sensory and nervous and directional control. In this descriptive-sectional study, 50 patients were studied in Primary Immune Deficiency. Based on the age of the patients, Audio tympanometry was taken from patients older than 4 years old and ABR and OAE were taken from younger patients. Thus, the presence or absence and the type of hearing loss were detected for patients. At The end, the abundance of these disorders, and their probable association with the type of Primary Immune Deficiency were studied. In a study of 50 patients, 32 (64%) of them were male and 18 (36%) of them were female. In the study of these patients, 13 (26%) of them had cellular primary immune deficiency, 33 (66%) of them had Primary humoral immune deficiency, and 4 (8%) of them had phagocytic primary immune deficiency. In the study of auditory status of the patients, 37 (74%) of them were with normal auditory and 13 (26%) of them had a variety of auditory abnormalities. From 13 patients with hearing loss, 10 (76.92%) patients were with conductive hearing loss, and 3 patients (23.7%) had a sensory neural hearing loss. From 13 patients listed, 2 (15.38%) patients had unilateral auditory disorder and in both cases the type of hearing loss was conductive. The hearing loss in 11 (84, 62%) of the remaining patients was bilateral. From 13 patients with disrupted hearing status in audiometric tests, 9 (69, 23%) of them were with primary humoral immunodeficiency. Based on the results of this study, hearing loss has a high prevalence in patients with Primary Immune Deficiency. Therefore, the extent of these abnormalities has a high prevalence in patients with primary humoral immune deficiency.

Introduction

In patients with immune defects, frequency of lower respiratory tract infections is a cause of chronic lung disease. The risk of recurrent otitis media is one of the cases occur in patients with immune deficiency. The incidence of these diseases may cause hearing defects in people that cause immune deficiencies are prone to recurrent infections of the middle ear. In some studies, the combination of safety defects with sensor neural hearing defects, conduction is discussed.

Mechanisms for sensor neural hearing deficits navigation safety defects can be expected to increase the risk of middle ear infections in patients with immune deficiencies are.

According to the articles review was carried out in cooperation with the hearing impaired immune deficiency, a comprehensive study has been done in this case. Also, early diagnosis of hearing loss in these patients at the right time can help patients to immune deficiency despite the evolutionary process auditory and speech training is troubled.

The aim of this study of hearing disorders in patients with primary immune deficiency is a hearing test (audio Tympanometry, ABR, OAE) patterns of sensory-neural hearing loss is conductive and sensory - nerve - conduction floor are classified.

Methods and Materials

This descriptive - cross - sectional. In this study, 50 patients with primary immunodeficiency referred to Children's Hospital in Tabriz and in Tabriz- Golgasht clinic during two years (beginning of 1392 to the end of 1393) were enrolled in full number. The sample size of the study

sample size estimation using the same studies for the study was calculated as follows: $N = Z^2 P (1-P) / d^2$

Inclusion criteria

Primary immunodeficiency disease
Parental consent for participation in the study.

Exclusion criteria

Patients with a precedent of more than 5 days in NICU ventilator Contact
Patients with a precedent of blood transfusions Hyperbilirubinemia
Patients with concomitant systemic diseases
Patients with a history of drugs auto Toxic
Patients with familial hearing loss
Patients with a history of low birth Apgar

In this study, 50 patients with primary immunodeficiency, primary immunodeficiency patients diagnosed with the same form that was designed to enter. Then, based on patients' age and cooperation with specialist ear, nose and throat patients over 4 years audio Tympanometry and patients younger than 4 years were ABR and OAE. Thus, the presence or absence and type of hearing loss was found for patients.

Finally, the prevalence of these disorders and the possible association with the type of primary immune deficiency were studied.

At baseline, all patients in the study were explained to the parents and patients were recruited with the consent of parents. For patients didn't impose any additional cost during the study. As well as any damage and loss of life for patients in the project did not exist. Patients participating in the study were free to leave the study at any time of the study. Please note that all information is kept strictly confidential patient records.

Result and Discussion

The survey collected data on 50 patients. Out of 50 patients, 32 patients (64%) were male and 18 patients (36%) were female. The average age of 72.14 ± 7.71 months in all patients studied (Min = 8, Max = 264), respectively. The average age of the male patients, 85.65 ± 11.14 (Min = 8, Max = 264) a month and in females, 50.11 ± 6.7 (Min = 10, Max = 108) months. The statistically significant difference in mean age between the sexes, with $P = 0.007$, respectively.

In this study, a family history of hearing loss, history of auto toxic medication, history of jaundice, and blood transfusions, history and history of hospitalization in NICU with a low Apgar score in any of the 50 patients there.

50 patients in the study, 13 patients (26%) with primary immunodeficiency cell, 33 patients (66%) with primary humoral immunodeficiency and 4 patients (8%) had a primary immunodeficiency phagocytic.

The survey of hearing patients, 37 patients (74%) had normal hearing and 13 patients (26%) have had some form of hearing loss. Of the 13 patients with hearing loss, 10 patients (76.92%) and 3 patients with conductive hearing loss (23.7%) have had sensorineural hearing loss. Of the 13 patients mentioned hearing loss in 2 patients (15.38%) in a way that in both cases, the hearing loss was the kind of guidance. The hearing impairment in the remaining 11 patients (84.62%) bilaterally.

In this study of 50 patients, 32 patients (64%) were male and 18 patients (36%) were female. In this study, a family history of hearing loss, prescription drugs auto toxic history, history of jaundice, and blood

transfusions, history and history of hospitalization in NICU with a low Apgar score in any of the 50 patients there. The other factors affecting the hearing of patients as much as possible have been removed.

Chandrasekhar and his colleagues study noted that a third of patients with positive safety defects, complaints or have the phone. The results of our study confirm these reports are. In our study of 50 patients with immune deficiency has been confirmed in 13 patients (26%) were varying degrees of hearing loss.

In a study of 25 patients by Berlucchi and his colleagues, they studied 18 patients with sensorineural hearing loss are conductive. This defect in 12 patients and bilateral in 6 cases were unilateral. The study also suggested that due to the significant prevalence of hearing impairment in patients with immune deficiencies, assess routine hearing tests performed in these patients.

In our study of 13 patients listed with hearing loss, 10 patients (76.92%) and 3 patients with conductive hearing loss (23.7%) have had sensorineural hearing loss. Also mentioned the 13 patients, hearing loss in 2 patients (15.38%) in a way that in both cases, the type of conductive hearing loss, which is in line with the results Berlucchi and colleagues.

In our study, hearing loss in the remaining 11 patients, 13 patients (84.62%), bilaterally. The high number of patients have bilateral disease is confirmed for early detection and management of patients with primary immunodeficiency hearing. Looking at the average age of the patients , we know that most of these patients are in the age range of most incoming data and learning with the help of interfaces is blind and deaf. Hearing impaired children in this

time, not only to learn they will drop off, but the isolation and loss of quality of life that is tangible.

In a study by Notarangelo and colleagues noted that antibody deficiency, the most common type of primary immunodeficiency. In our study of 50 patients, 13 patients (26%) with primary immunodeficiency cell, 33 patients (66%) with primary humoral immunodeficiency and 4 patients (8%) had a primary immunodeficiency phagocytic.

Notarangelo and colleagues reported in a study that the most common form of primary immunodeficiency humoral immunodeficiency that clinical presentation with recurrent infections of the ear, throat and nose. The results obtained in our study are in support of this. In our study, of 13 patients with hearing impairment tests audiometry, 9 patients (69.23%) with primary humoral immunodeficiency, respectively.

Polmar and colleagues noted in a study that most patients with antibody deficiency, recurrent infections of the ear, nose and throat may be the only sign of immunodeficiency and should be considered. Therefore, given that the most common form of primary immunodeficiency humoral immunodeficiency and the prevalence of hearing loss in these patients, the diagnostic procedures for these children is determined:

Periodic review of the hearing of patients with primary immunodeficiency (especially in patients with primary humoral immunodeficiency)

Evaluation of all patients with a history of recurrent infections and chronic ENT and lack of functional and structural defects in humoral immunity in the upper respiratory tract.

For recurrent infections and chronic ear, nose and throat is one of the most fundamental problems the health system is in the form of a clear, quality of life in adults and children, although proper diagnosis and treatment are affected. Although accurate diagnosis and proper treatment, groups of these patients do not respond to conventional therapy and the symptoms are similar, so check the status of the immune system (humoral) of these patients is necessary.

Measurement of physiological functions such as the cochlea and auditory nerve otoacoustic emission and auditory brainstem response in differentiating between hearing loss and impaired cochlear damage receivers helpful.

Otoacoustic emissions, cochlear outer hair cells produce sounds that are weak and in normal people by placing a microphone in the external auditory canal in response to sound stimulation or transient distortion results are recorded. The origin of cochlear hearing loss, OAEs evoked by sound, absent or reduced.

Auditory brainstem response in the auditory pathway are produced by the structure of the waves I and II, from the auditory nerve activity and wave III, IV and V from the structures of the auditory brainstem. ABR wave latencies between nerve signal transmission speeds. Beyond the cochlear nerve disorders may reduce the transmission rate and thus increases the latency between the ABR waves.

Severe ABR impairment (two standard deviations above normal values) or not observed in the record OAEs, auditory nerve dysfunction and normal functioning hair cells in the cochlea of the team. The results of this study have a high prevalence of

hearing loss in patients with primary immunodeficiency is. Of those, the primary humoral immunodeficiency in patients with these disorders is very high prevalence. Regarding the relationship between hearing impairment and primary immunodeficiency, routine evaluation of patients with immune deficiency known of auditory function is recommended in the review period. It also evaluated in patients with primary immunodeficiency humoral within shorter and more should be done carefully.

Also, on the contrary, all patients with a history of recurrent infections and chronic ENT and lack of functional and structural defects specially in humoral immunity upper respiratory tract should be evaluated.

Conclusion

Based on the results of this study, hearing loss has a high prevalence in patients with Primary Immune Deficiency. Therefore, the extent of these abnormalities has a high prevalence in patients with primary humoral immune deficiency.

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