Is hearing loss overlooked in patients with hepatic glycogen storage diseases?

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Hepatic glycogen storage diseases are a group of disease manifesting mainly with hypoglycemia and hepatomegaly. The patients require frequent daytime and nocturnal feedings. Hypoglycemia may cause sensorineural hearing loss and nocturnal feeding is a risk factor for development of gastroesophageal reflux that may cause chronic otitis media and hearing loss consequently. We aimed to determine the prevalence and characteristics of hearing loss in hepatic glycogen storage diseases.

A total of 24 patients with hepatic glycogen storage disease (15 glycogen storage disease type I and 9 non type I) and 24 age/sex matched healthy controls were enrolled in the study. Pure tone audiometer, immittance, acoustic reflex measurement, otoacoustic emission test (OAE) and auditory brainstem response (ABR) tests were applied to all participants.
Introduction and methods

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Results

- Hearing loss was determined in 17/24 patients (12 glycogen storage disease type I and 5 non type I) with pure tone audiometer.
- 8 patients had slight and 9 patients had mild hearing loss.
- 14 patients had type C tympanogram and middle ear pressures were significantly decreased in patient group.
Results II

• I-V interpeak latencies were prolonged in 9 (37.5%) patients. Since, these patients also had type C tympanograms, they were diagnosed with mixed HL.

• Interpretation of all of the audiologic findings determined that 9 patients had mixed type and 8 had conductive type hearing loss.
Summary

• Conductive/mixed hearing loss suggests chronic middle ear dysfunction.

• The cause of mixed hearing loss might be associated with fluid collection in the middle ear as a result of GER due to nocturnal feeding, feeding in supine position and presence of more horizontal eustachian tube in children.

• Non type I and type I patients >12 years old were on less frequent nocturnal feeding and type I patients > 12 years old had normally located eustachian tube. In these patients, prevalence of abnormal middle ear pressures and hearing loss were lower than type I patients <12 years old.

• To the best of our knowledge this the first study to comprehensively assess auditory functions in hepatic GSD patients comparing with type I and non type I patients.

• Even in slight/mild HL, decreased academic performance, social and speech development were reported many times. Therfore routine hearing assessment may be recommended in hepatic GSD patients to prevent long term complications.

• We suggest the underlying cause of the hearing loss in the patients may be associated with gastroesophageal reflux beyond hypoglycemia, however further studies in large number of patients with the evaluation of presence of reflux are required to enlighten the pathogenesis.