

# Unilateral hyperacusis in a normal-hearing child

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There is some debate on the epidemiological data of childhood tinnitus and hyperacusis. It is vital for Audiologists to pay careful attention to minor complaints such as hyperacusis in children to manage them successfully.

## Keywords:

childhood, hyperacusis, phonophobia, speech-evoked auditory brainstem response

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## Introduction

Hyperacusis is often present in association with tinnitus. Jastreboff and Hazell [1] defined hyperacusis as an abnormal sound sensitivity that arises from the auditory system, either peripheral or central. They suggest that decreased sound tolerance consists not only of hyperacusis but it also a fear of sound (phonophobia) or a strong dislike of sound (misophonia). They also reported that patients with misophonia or phonophobia experience abnormally strong sensitivity of the limbic and autonomic nervous system without involvement of the auditory system.

Prevalence studies of childhood hyperacusis are scarce. Coelho *et al.* [2] found a prevalence of 3.2% using their previously introduced definition on a population of 506 children, age 5–12 years.

## Case report

A 5-year-old boy was referred from the Ear, Nose and Throat (ENT) Outpatient Clinic, Demerdash Hospital, Ain Shams University, with reported pain and intolerance of loud sounds in the left ear that started 3 months ago. He denied any other ear symptoms and had never been operated on in this ear before. He had no psychiatric comorbidities, and had normal developmental history and past history (prenatal, natal, and postnatal).

Further investigations were performed to rule out causes of hyperacusis. A thorough ENT clinical examination and otological and neurological assessments showed normal findings. He subsequently underwent audiological tests that (pure-tone audiometry, speech discrimination score, tympanometry, and the stapedius reflex were normal (Fig. 1).

TEOAEs indicated a bilateral pass response. The click-evoked auditory brainstem response also showed no abnormality (Fig. 2).

Further auditory brainstem assessment was performed to uncover any subtle dysfunction at higher brainstem levels by speech-evoked auditory brainstem response (SABR). Surprisingly, he had completely normal SABR in the right ear in the form of repeatable waves with normal latencies and amplitudes, matching the BIOMARK age norms. However, the waves were highly distorted and unrepeatable in the left ear even after repeating the test 1 week later (Fig. 3).

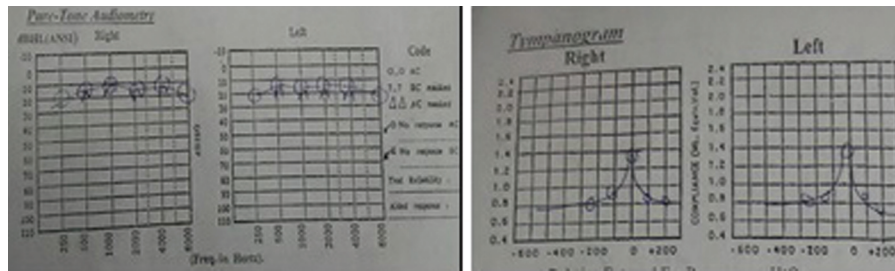
MRI of his skull indicated no lesions in the brain, pons, or the auditory pathway.

One month later, his mother visited our clinic again reporting that 1 week ago, her child had an attack of severe headache along the left side of the head associated with abdominal pain, vomiting, and diarrhea. The mother believed that this was gastroenteritis, but she noticed that the attack subsided after sleep and did not seek any medical advice till the child experienced the same attack 2 days before the visit. During this visit, the SABR test was repeated and the results showed the same pattern. The child was diagnosed with a probable history of migraine. On consultation with the pediatric neurologist, avoidance of migraine triggers and trial of preventive medical treatment for migraine were started for 3 months (in the form of Topamax (Janssen Pharmaceuticals, a Johnson & Johnson subsidiary) 25 mg tablets; one tablet was crushed and mixed with about two tablespoons of fruit juice or soft food as yogurt, honey, or jam).

Re-evaluation was carried out after 3 months using the only abnormal test in the previous evaluations, which is SABR. Surprisingly, left ear waves had improved (Fig. 4). Moreover, the child reported relief of pain and better tolerance to loud sounds in the left ear.

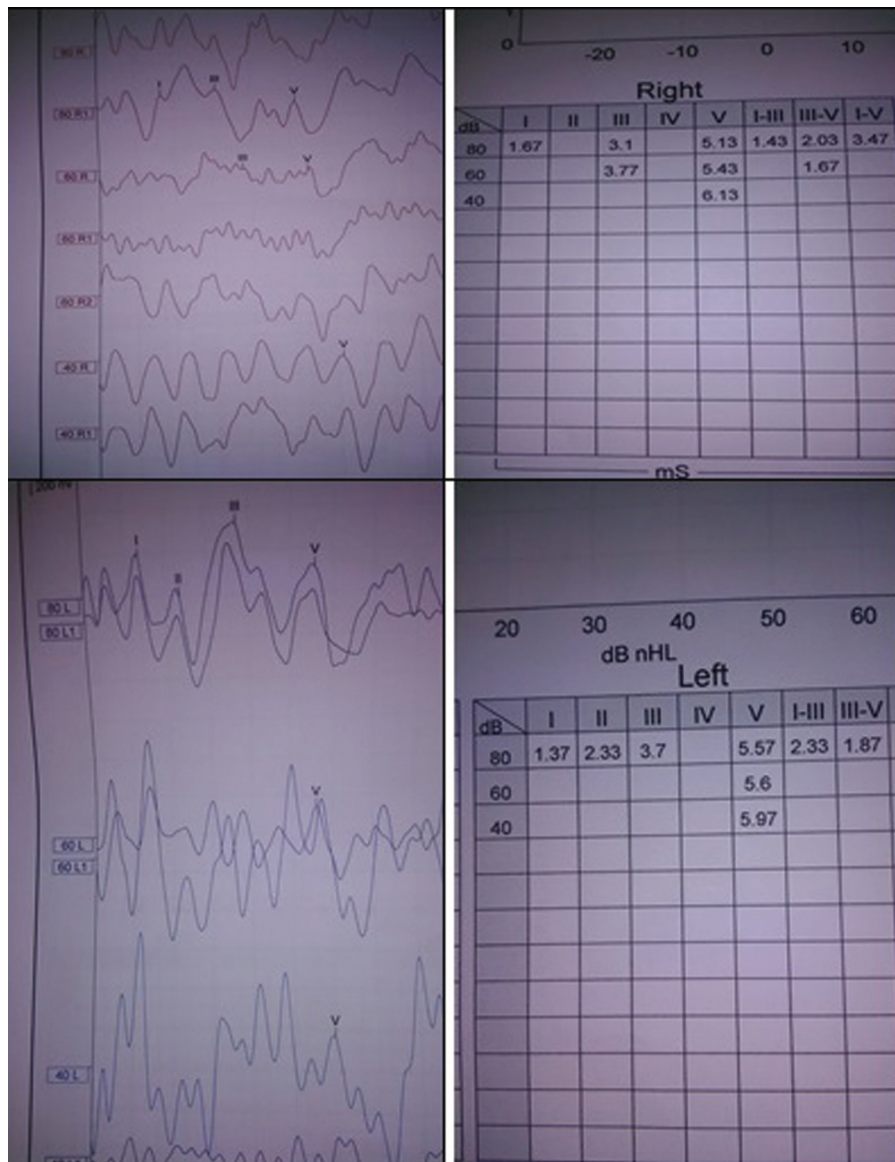
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Figure 1



Pure-tone audiometry and tympanometry.

Figure 2



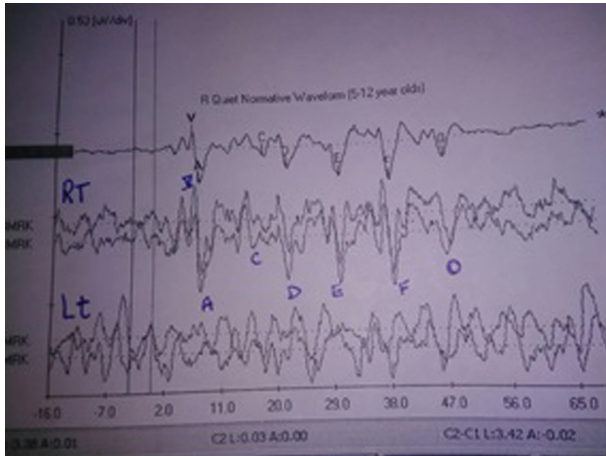
Click-evoked auditory brainstem response in both ears (the child was awake).

**Discussion**

Clinically, hyperacusis can be caused by lesions in the peripheral or central auditory system [3]. Central causes can be migraine, depression, head injury,

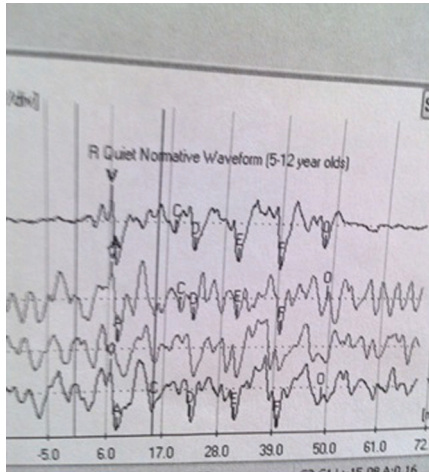
William’s syndrome, or multiple sclerosis. Patients with hyperacusis or phonophobia may first seek treatment in the general practitioner’s clinic or general outpatient clinics, and these doctors usually

Figure 3



Speech-evoked auditory brainstem response in both ears using speech syllable 'da' 40 ms.

Figure 4



Speech-evoked auditory brainstem response in the left ear after 3 months of medical Treatment (TTT).

then refer the patient to specialized clinics (e.g. the ENT, psychiatry, or neurology or the general physician's clinic according to the suspected diagnosis). Thus, assessment of the medical history is vital at the first consultation. The patient should be investigated carefully to rule out serious causes of hyperacusis. Objective audiological assessments are among the tests that can be performed, including the acoustic reflexes and the auditory-evoked potentials, in addition to MRIs to rule out peripheral and central causes within the auditory system [2,4,5].

In the present case, all these tests were negative, except SABR in the left ear, reflecting affection of the higher brainstem functions. MRI testing is expensive and not widely available, but it was ordered in this case after unilateral abnormality was recorded in the SABR test to

rule out serious central causes of hyperacusis. However, it should not be a routinely performed test here.

In practice, most patients with hyperacusis improve after determining the causative disease and treating it. Baguley and Andersson [6], in their latest book on hyperacusis, suggest that 'addressing hyperacusis must always involve the classical auditory system as well as systems of emotion and behavior – and as such is both physiological and psychological at the same time'. In many cases, their statement is valid.

Classic symptoms of migraine include severe throbbing headache on one side of the head, nausea, and vomiting and extreme sensitivity to light and noise. Although these symptoms seem common to patients with migraine, there is also an association between migraine and the inner ear and brain mechanisms that may influence hearing and balance.

Vestibular symptoms of migraine are vertigo, imbalance, dizziness, unsteadiness, and extreme sensitivity to motion.

Hearing symptoms include ear fullness and tinnitus or ringing in the ear. The hearing symptoms are associated with vertebrobasilar migraine.

The cause of migraine headaches probably relates to both abnormal discharges in cells within the brain and to the constriction of the walls of the blood vessels in and around the brain. Although migraine can be associated with inner ear balance and mechanisms, symptoms of migraine can appear independent of headaches.

Topiramate (TPM) is an oral drug that is used to prevent the seizures of epilepsy. It is an antiepileptic drug. It also prevents migraine headaches. Although the exact mechanism of action of TPM is unknown, scientific studies suggest that it may alter neurotransmitters within the brain. By altering the production or action of the neurotransmitters, TPM may suppress the abnormal activity of the nerves in the brain that cause the seizures or may prevent the abnormal activity from spreading to other nerves. Other studies suggest that TPM may suppress the nerves directly (i.e. not by altering neurotransmitters and make them less likely to fire. The negative cognitive effects and weight gain induced by TPM often are temporary and resolve after medication discontinuation [7].

In conclusion, hyperacusis involves audiological, emotional, and behavioral components, and the majority of patients may first present to outpatient clinics or general practitioners. Thus, the attending doctors should include

SABR testing in the diagnosis of hyperacusis for its successful management.

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#### Conflicts of interest

There are no conflicts of interest.

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