ISSN: 2083-389X
eISSN: 2084-3127

© J Hear Sci, 2022; 12(2): 63–65
DOI: 10.17430/JHS.2022.12.2.6

STATIC ENCEPHALOPATHY AND SENSORINEURAL HEARING LOSS: A SINGLE CASE STUDY

Contributions:
A Study design/planning
B Data collection/entry
C Data analysis/statistics
D Data interpretation
E Preparation of manuscript
F Literature analysis/search
G Funds collection

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Abstract

Introduction: The term static encephalopathy is a phrase used by neurologists for chronic nonprogressive brain disorders in children, primarily cerebral palsy and mental retardation. As defined by Seals, static encephalopathy is permanent or unchanging brain damage. Depending on the severity of the damage and the part of the brain involved, development changes can vary. The aim of the present study was to use multiple tests, including behavioural, physiological, electrophysiological, and radiological evaluations, to analyse the hearing of a case with static encephalopathy.

Case report: The current study describes a 3 year old male child who was brought in with a complaint of having reduced hearing sensitivity in both ears for more than 1 year. The subject had no family or medical history which could directly or indirectly have an impact on auditory system, directly or indirectly. Previous audiological evaluation had revealed bilateral severe to profound hearing loss. Radiological evaluation revealed structural abnormalities.

Conclusions: The patient presented in this case study went through multiple tests including behavioural, physiological, electrophysiological, and radiological. All the evaluations point to the conclusion that static encephalopathy can produce sensorineural hearing loss.

Key words: static encephalopathy • physiological measures • electrophysiological measures

ENCEFALOPATIA STATYCZNA A NIEDOSŁUCH ODBIORCZY: STUDIUM PRZYPADKU

Streszczenie

Wprowadzenie: Pojęcie encefalopatii statycznej jest stosowane przez neurologów do opisu przewlekłych, niepostępujących zaburzeń centralnych u dzieci, przede wszystkim porażenia mózgowego i upośledzenia umysłowego. Zgodnie z definicją organizacji Easter Seals encefalopatia statyczna jest to stałe i nieulegające zmianom uszkodzenie mózgu. Zmiany rozwojowe są różne w zależności od stopnia uszkodzenia i tego, jakiej części mózgu ono dotyczy. Celem pracy było zastosowanie wielu badań, w tym: behawioralnych, fizjologicznych, elektrofizjologicznych i radiologicznych, do analizy słuchu pacjenta z encefalopatią statyczną.

Opis przypadku: Badanie opisuje przypadek 3-letniego dziecka płci męskiej, który został przywieziony ze zgłoszeniem obniżonej czułości słuchu w obojgu uszach utrzymującej się od ponad roku. Pacjent nie miał ani obciążonej historii rodzinnej, ani medycznej, która mogłaby mieć bezpośredni lub pośredni wpływ na układ słuchowy. Wcześniejsze badanie audiologiczne wykazało obustronny niedosłuch stopnia znacznego do głębokiego. W badaniu radiologicznym widoczne były nieprawidłowości anatomiczne.

Wnioski: Pacjent, którego przypadek omawiamy, przeszedł szereg badań, w tym badania: behawioralne, fizjologiczne, elektrofizjologiczne i radiologiczne. Otrzymane wyniki prowadzą do wniosku, że encefalopatia statyczna może powodować niedosłuch odbiorczy.

Słowa kluczowe: encefalopatia statyczna • pomiary fizjologiczne • pomiary elektrofizjologiczne

Introduction

Encephalopathy means any disorder or disease of the brain, especially chronic degenerative conditions [1]. In modern usage, encephalopathy does not refer to a single disease, but rather to a syndrome of overall brain dysfunction; this syndrome has many possible organic and inorganic causes. The term static encephalopathy (SE) is used to refer to chronic non-progressive brain disorders, as in children with cerebral palsy and mental retardation [2]. As defined by Seals, SE is permanent or unchanging brain damage. Depending on the severity of the damage and the part of brain involved, development changes vary. Developmental problems can include a range of disabilities such as cerebral palsy, learning disabilities, mental retardation, autism, speech delays, attention deficits, hearing and vision impairments, and oral motor problems. Every child with static encephalopathy is unique [3]. SE of childhood with neurodegeneration in

adulthood (SENDA) is a recently established disorder that is a subtype of neurodegeneration with brain iron accumulation (NBIA) [4]. In high-income countries, neonatal encephalopathy occurs in 1 to 3 per 1000 live births; approximately 20–25% of affected infants die, and 40% of survivors have significant brain injury and lifelong disability [5–7]. It is reported that permanent hearing loss of all degrees of severity can occur in grades of hypoxic ischaemic encephalopathy (HIE). They also indicate that auditory neuropathy spectrum disorder can develop secondary to HIE.

The degree of deficits in an individual with SE depends on the location and extent of the underlying CNS injury. The site of lesion and underlying cause can often be predicted based on the clinical pattern. However, the deficits of most individuals with SE remain of uncertain pathogenesis, although rapid progress in the elucidation of genetically determined developmental disorders is reducing this uncertainty and replacing these generic labels with specific conditions. The importance of labels is that they can designate a definite cause. Increasing specificity of labels makes it possible to replace the generic label of SE with terms better relating to heritability, risks, prognosis, and treatment.

Some of the neurological characteristics of SE include: easily distracted and disorganized; poor judgment; difficulty with abstract maths and money management; difficulty with short term memory; emotional immaturity; and inability to control impulses.

The aim of the present study was to use multiple tests including behavioural, physiological, electrophysiological, and radiological tests to analyse the hearing of a case with static encephalopathy.

Case report

A 3-year-old male child was brought to the clinic with a complaint of reduced hearing sensitivity in both ears since age 1–2 years. There was no significant prenatal or perinatal medical history or family history. However, the postnatal medical history revealed paroxysmal events at 2 months of age with semiology showing tonic extension of the neck with rolling up of the eyes. Earlier evaluations had included audiological and radiological (MRI) examinations which had pointed to bilateral severe to profound hearing loss.

Method

A detailed case history was obtained. Otoscopic evaluation visualized the external auditory canal and tympanic membrane. All audiological tests were carried out in a sound-treated room. Tympanograms and acoustic reflex thresholds were assessed using a GSI Tympstar. Tympanogram was measured using a 226 Hz probe tone, with pressure sweep from −200 to +400 daPa. Ipsilateral and contralateral acoustic reflexes were obtained using pure tone stimuli of 0.5, 1, 2, and 4 kHz at 90, 100, and 110 dB. DPOAEs were recorded using IHS Jr instrument to check outer hair cell (OHC) function using an F2/F1 ratio of 1.2 and tone pairs of 65 and 55 dB SPL for L1 and L2, respectively. Two separate runs per ear were collected

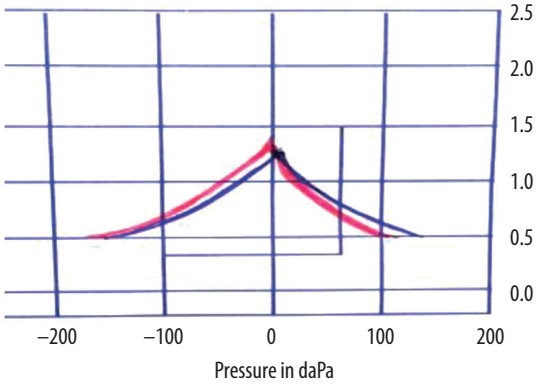

Figure 1. Tympanometry findings for both right (red) and left (blue) ear

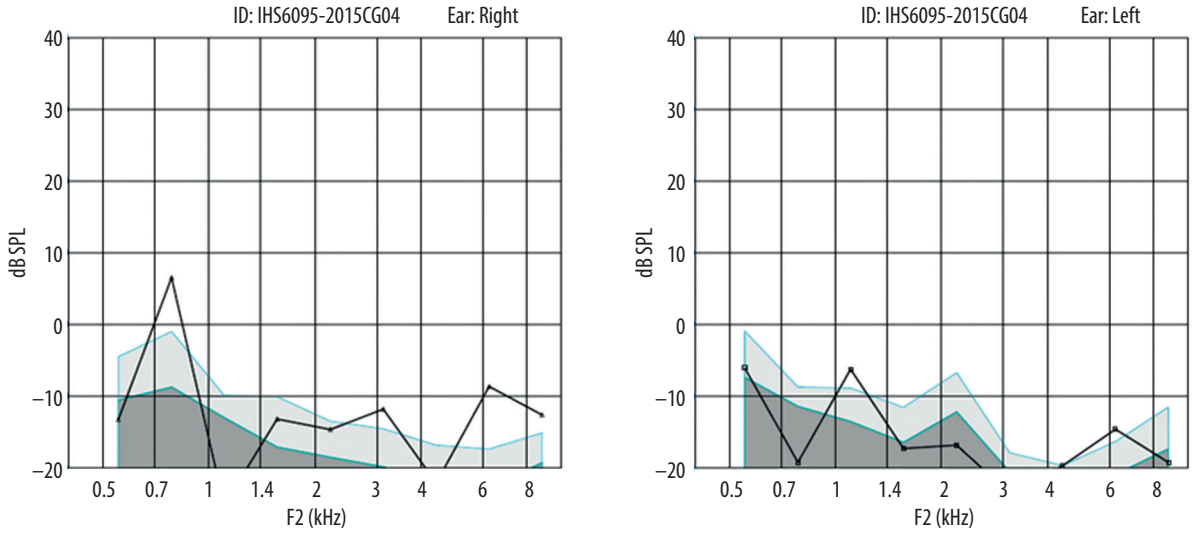

Figure 2. DPOAE findings for both right and left ears

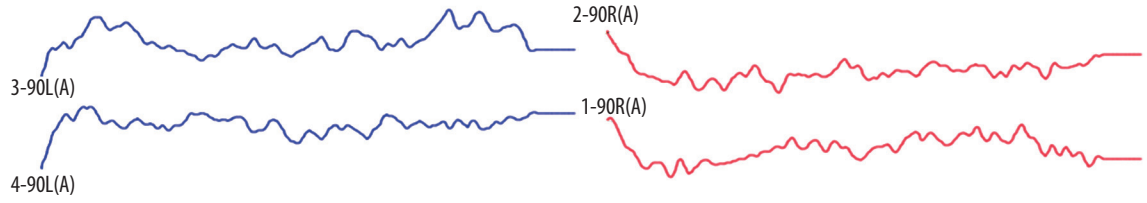

Figure 3. Auditory brainstem responses for right (red) and left (blue) ears

to determine repeatability. Auditory brainstem responses (ABRs) were done using the IHS Jr instrument using 1 ms clicks and recorded in a 10 ms time window using a band-pass filter from 30 to 3000 Hz. Rarefaction clicks were presented at a rate of 11.1 per second in a quiet environment. Horizontal placement was used using surface electrodes and the electrode impedance was regularly monitored.

Results and discussion

Physiological evaluation: Tympanometry revealed bilateral "A" type tympanogram (**Figure 1**) suggestive of normal middle ear functioning. Reflexometry revealed bilateral absence of ipsilateral and contralateral reflexes.

OAE evaluation (**Figure 2**) showed bilateral absence of DPOAEs suggestive of outer hair cell dysfunction.

Electrophysiological evaluation: ABR results (**Figure 3**) revealed bilateral absence of wave V at 90 dBnHL, suggesting severe to profound hearing loss.

Conclusions

The patient presented in this case study had gone through multiple tests including behavioural, physiological, electrophysiological, and radiological. All the evaluations point to the conclusion that static encephalopathy can lead to sensorineural hearing loss [8]. It has been reported that sensorineural hearing loss is frequent in cases of cerebral palsy. Early audiological assessment is important to improve the language outcome in these children.

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