

# UNUSUAL MAGNETIC RESONANCE FINDINGS IN TWO CHILDREN WITH SUDDEN SENSORINEURAL HEARING LOSS

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**ABSTRACT - Objective:** To describe the MRI findings of two pediatric patients with sudden sensorineural hearing loss (SSHL). **Case reports:** Two male patients (two-year and three-months-old, and one year and four-months-old) presented with sudden dumbness. Physical and neurological examinations were unremarkable besides bilateral hypoacusia. All the laboratory investigation was negative, and brain stem auditory evoked potentials showed deep bilateral deafness in both cases. MRI studies revealed normal inner ears and multifocal white matter areas of slight low signal on T1-weighted images and high signal on FLAIR images. The follow-up MRI studies and neurological examinations did not demonstrate alterations in the previous findings. **Conclusion:** Pediatric patients with SSLH may present cerebral white matter signal abnormalities at the MRI as the only finding. Further studies with larger casuistics need to be conducted to elucidate these findings.

**KEY WORDS:** magnetic resonance imaging, sudden sensorineural hearing loss, cerebral white matter.

## Achados incomuns nas ressonâncias magnéticas de duas crianças com perda auditiva neuro-sensorial súbita

**RESUMO - Objetivo:** Descrever os achados de ressonância magnética (RM) em dois pacientes pediátricos com perda auditiva neuro-sensorial súbita (PANS). **Relato dos casos:** Dois pacientes masculinos (dois anos e três meses e um ano e quatro meses de idade) apresentaram-se com surdez súbita. Os exames físico e neurológico foram normais, à exceção da hipoacusia. Toda a investigação laboratorial foi normal, e os potenciais auditivos evocados do tronco cerebral demonstraram surdez profunda bilateral. Os exames de RM evidenciaram estruturas normais dos ouvidos internos e áreas multifocais de discreto hipossinal em T1 e hipersinal em FLAIR na substância branca dos hemisférios cerebrais. As RMs e os exames neurológicos de controle não demonstraram alterações nos achados prévios. **Conclusão:** Pacientes pediátricos com PANS podem apresentar à RM alterações de sinal na substância branca dos hemisférios cerebrais como único achado. Estudos futuros com casuísticas maiores deverão ser conduzidos para a elucidação destes achados.

**PALAVRAS-CHAVE:** ressonância magnética, perda auditiva neuro-sensorial súbita, substância branca cerebral.

Sudden sensorineural hearing loss (SSHL) is defined as a sensorineural hearing loss with at least 30-dB decrease in threshold in three contiguous test frequencies occurring over a 24 to 72-hour period<sup>1</sup>. This condition is usually unilateral and may be complete<sup>2</sup>. Many different causes of SSLH have been hypothesized, such as viral infection of the labyrinth or cochlear nerve, vascular insult, perilymphatic hypoxia, intralabyrinthine membrane rupture, inflammatory and metabolic causes, immune-mediated inner ear disease, perilymphatic fistulas and others<sup>3,4</sup>.

Viral infection is the most common presumed

cause of SSLH<sup>2,5</sup>. Nearly one-third of the patients with SSLH has abnormal MRI findings, generally labyrinth or internal auditory alterations<sup>4,6</sup>. However, isolated white matter signal alteration at the MRI was not previously reported in patients with SSLH.

We present two pediatric patients with SSLH who had at the brain MRI diffuse alteration of the white matter signal.

## CASE

The study was approved by the ethical committee of our hospital and the parents signed informed consent.

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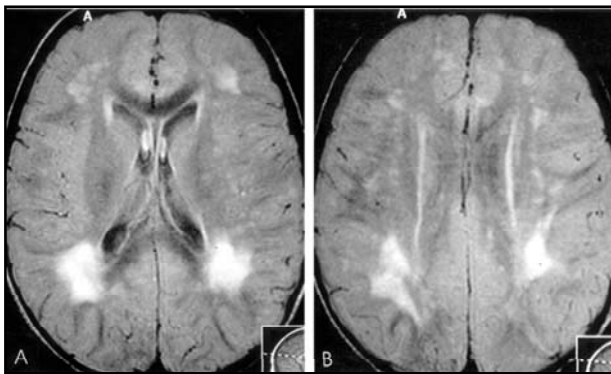
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**Case 1** – A two-year and three-months-old male patient was seen for sudden dumbness. At the time of examination the child was monosyllabic and hypoacusia was observed. The mother referred a pregnancy and delivery with no intercorrences. She also mentioned a discrete psychomotor delay and irritable behavior. During lactation period the child had repetitive otitis. Physical examination showed only global hypo-reflexia and hypoacusia.

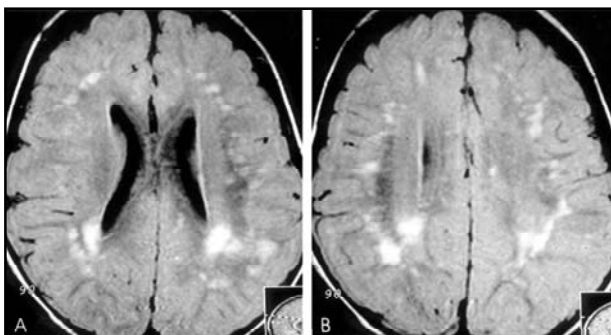
Laboratory tests were performed resulting negative immune tests for toxoplasmosis, syphilis, HIV, CMV and rubella. Biochemical tests and hemogram were all normal. A brain stem auditory evoked potential with stimulation clicks from 1000Hz to 4000Hz showed no eletrophysiological potentials at both sides, demonstrating a deep bilateral deafness.

The CT-scan demonstrated multifocal hypodensities in the white matter, mainly at the posterior regions. The MRI (1.5T GE LX echo-speed plus - General Eletric Medical Systems, Milwaukee, WIS, USA) was performed with routine sequences (T1, T2, FLAIR, T2\*, diffusion and post-contrast T1). This exam revealed a normal and symmetric configuration of the inner ears and multifocal white matter areas of slight low signal on T1-weighted images and high signal on FLAIR images (Fig 1). The other MRI sequences showed no additional findings.

In the one-year follow-up the patient persisted with severe deafness and the same neurological examination and neuroimaging findings.



*Fig 1. A and B. FLAIR MRI images show multifocal areas of high signal in the white matter, mainly in the posterior regions.*



*Fig 2. A and B. FLAIR MRI images demonstrates multifocal areas of high signal in the white matter.*

**Case 2** – A one year and four-months-old male patient was seen for sudden decrease of speech fluency and unstable gait, the last one for a few days only. At this time the auditory evoked potential show a loss of 30% of audition on both ears. As a past history, at eleven months-old he had febrile illness of unknown etiology. Physical and neurological examinations were unremarkable besides bilateral hypoacusia.

Laboratory studies showed negative tests for toxoplasmosis, syphilis, HIV, CMV and rubella. Biochemical tests and hemogram were normal. Nerve conduction studies and thyroid function were also normal. A brain stem auditory evoked potential with stimulation clicks from 1000 Hz to 4000 Hz showed no eletrophysiologic potential at both sides, demonstrating a deep bilateral deafness. Fundoscopic examination was unremarkable.

The head CT-scan revealed multifocal hypodensities in the white matter. The MRI (1.5T GE LX echo-speed plus -General Eletric Medical Systems, Milwaukee, WIS, USA) was performed with routine sequences (T1, T2, FLAIR, T2\*, diffusion and post-contrast T1). This exam showed no abnormalities in the inner ears and multifocal white matter areas of slight low signal on T1-weighted images and high signal on FLAIR images (Fig 2). The other MRI sequences showed no additional findings.

In the two-year follow-up he persisted with profound deafness and the same neurological examination and MRI findings.

## DISCUSSION

The sensorineural hearing loss (SHL) occurs when the acoustic pathway between the oval window and the auditory cortex is disrupted. It can be subdivided into sensorial (cochlear) and neural (retro-cochlear) components. The neural hearing loss occurs when a lesion affects more proximal elements of the acoustic pathway, including the vestibulocochlear nerve, brain stem or midbrain nuclei, auditory tracts or superior temporal gyrus<sup>7</sup>. The imaging evaluation of these patients generally includes an analysis of the acoustic pathway, from the cochlea to the auditory cortex and the MRI is the modality of choice for this patients<sup>8</sup>. The main causes of SHL are neoplasms (acoustic neuromas and meningiomas), congenital abnormalities, infections, autoimmune processes, circulatory and metabolic abnormalities, trauma, intrauterine exposures and perinatal insults and degenerative and idiopathic conditions<sup>2</sup>. From all this causes, the genetic anomalies, with or without associated hereditary syndromes, cause approximately half of the cases of profound childhood deafness<sup>2</sup>. Intrauterine exposures and perinatal insults account for many other cases of childhood sensorineural hearing loss, and the

cause remains unknown in 25-40% of cases<sup>2</sup>. Our patients had no familiar history of hereditary syndromes nor intrauterine exposure to infections (negative laboratory tests).

When the sudden sensorineural hearing loss is considered, the main cause is no longer genetic, but idiopathic, although many studies indicate a probable viral etiology<sup>2-5</sup>. The viral SSHL usually is unilateral<sup>2,3</sup>, and compromises only the labyrinth and/or the cochlea, causing a labyrinth enhancement on MRI<sup>4</sup>. Other reported causes for SSHL also appear as labyrinth enhancement (immune labyrinthitis, syphilis, idiopathic) or internal ear anomalies (vascular anomalies and Meniere disease)<sup>4</sup>. The only reported cause that may present with diffuse white matter alteration on MRI is multiple sclerosis, but this entity is not characterized only by SHL<sup>9</sup> as seen on the cases presented, and it does not affect this age group. Our patients had bilateral SSHL and no evidence of labyrinth enhancement or internal ear anomalies at the imaging studies.

There are a great number of diseases that may affect the white matter and may course with hearing loss. However these diseases generally have a lot of other characteristics other than the hearing loss, and none of them were seen in our cases. Some diseases that may presented with hearing loss as one of its symptoms are: mitochondrial diseases (chronic progressive external ophthalmoplegia; Kearns-Sayre syndrome)<sup>10</sup>, Susac syndrome (microangiopathy of the retina, cochlea and brain)<sup>11</sup>, vascular diseases, toxic demyelination and inherited demyelination diseases. All of these entities may have imaging findings similar to those of our cases, but all of them require another criteria (both clinical and radiological) to be diagnosed.

Another possibility for the cases reported is post-infectious encephalitis which may occur after

a specific viral disease, after an upper respiratory tract infection, after vaccination or spontaneously. This disease results in acute demyelination, generally focal, and has an ample variety of clinical manifestations, which may be transitory or permanent. This can be seen, for instance, in patients who had prenatal exposure to the rubella virus, causing usually hearing loss and psychomotor retardation. These patients may show periventricular and subcortical hyperintensities and delayed myelination on MRI study<sup>12</sup>.

In conclusion, although we are not certain about the cause of the abnormalities on the MRI of these cases, they can show us that patients who underwent MRI as a part of the investigation of SSHL may present white matter signal abnormalities as the only finding.

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