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Evaluation of the outcome of CT and MR imaging in pediatric patients with bilateral sensorineural hearing loss

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ABSTRACT

Objective: To evaluate the clinically relevant abnormalities as visualized on CT and MR imaging in children with symmetric and asymmetric bilateral sensorineural hearing loss (SNHL), in relation to age and the severity of hearing loss.
Study design: Retrospective cohort study.
Setting: Tertiary referral otology and audiology center.
Patients and diagnostic interventions: From January 2006 until January 2016, a total of 207 children diagnosed with symmetric and asymmetric bilateral SNHL were included. They underwent CT and/or MR imaging for the evaluation of the etiology of their hearing loss.
Main outcome measures: Radiologic abnormalities associated with SNHL.
Results: 302 scans were performed in 207 children (median age of 0.8 years old) with bilateral SNHL. The most frequently identified cause of bilateral SNHL was a malformation of the labyrinth. The combined diagnostic yield of CT and MR imaging was 32%. The diagnostic yield of MR (34%) was considerably higher than that of CT (20%). We found a higher rate of abnormalities in children with profound hearing loss (41%) compared to milder hearing loss (8–29%), and in asymmetric SNHL (52%) compared to symmetric SNHL (30%).
Conclusion: Imaging is essential in the etiologic evaluation of children with bilateral SNHL. The highest diagnostic yield is found in children with bilateral asymmetric SNHL or profound SNHL. Based on our findings, MR is the primary imaging modality of choice in the etiological evaluation of children with bilateral SNHL because of its high diagnostic yield.

1. Introduction

Sensorineural hearing loss (SNHL) in pediatric patients may be present at birth or become apparent later during infancy. In both cases, the cause of the hearing loss may be hereditary or acquired. Congenital hearing impairment is the most common birth defect, with an incidence of 1,9 in 1000 newborns in the Netherlands and 1 to 3 per 1000 births worldwide [1,2]. In the Netherlands, congenital hearing loss is detected at a very early age by the current newborn hearing screening program provided by the Dutch Child Health and Welfare service (JGZ) which was implemented from 2002 to 2006 [3]. Currently, 96,5% of the newborn in The Netherlands are screened for hearing impairment [1].

Screening for hearing loss during the newborn period has led to early detection and diagnosis of SNHL, facilitating timely intervention [4,5]. Congenital hearing loss is nowadays generally detected within the first weeks of life in The Netherlands, however hearing loss may also be diagnosed later during infancy, because of a late onset (due to acquired pathologies such as infection or trauma) or a progressive nature of hereditary etiologies.

Whereas adequate and timely revalidation has been the primary goal of newborn hearing screening, it has also sparked the interest in the causes of pediatric SNHL. In addition to genetic and laboratory

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testing, imaging by computed tomography (CT) and/or magnetic resonance (MR) imaging has become an essential part of the evaluation of pediatric SNHL [6–8]. CT and MR imaging are regarded as complementary modalities. CT is considered a better modality for the identification of bony abnormalities, while MR imaging provides superior information about the cochlear nerve, the intracranial structures and early stages of fibrosis in cases of meningitis [9]. Previous studies of children with SNHL show temporal bone abnormalities in 18–37% when CT is performed, or 24–33% when MR imaging is performed as a single modality [6,7]. Combined, the overall reported diagnostic yield is 25–38% [10,11].

Children with SNHL form a heterogeneous group of patients, with a varying age at detection and varying degrees of hearing loss. These different patient groups may represent different SNHL etiologies, resulting in a different radiologic outcome and yield. Here, we evaluate the prevalence and spectrum of causative radiological abnormalities in children with bilateral SNHL and their associations with the severity of the hearing loss, the symmetry of the hearing loss and the age at diagnosis.

2. Materials and methods

2.1. Patients

Patients between 0 and 18 years of age diagnosed with bilateral SNHL were referred for etiological evaluation to the VU University Medical Center (VUmc) in Amsterdam, The Netherlands. The majority of these children was referred directly after detection of their hearing loss by the Dutch newborn hearing screening, and subsequent bilateral SNHL was confirmed by the audiological center of the VUmc or other regional audiology centers. In some cases, the detection of SNHL or need for etiological evaluation arose later in life and referral took place at an older age, either by audiology centers, general practitioners, the Dutch child health and welfare service, or otorhinolaryngologists. At the VUmc, the etiological evaluation was performed by a dedicated multidisciplinary team (The Center for Diagnostics of Sensorineural hearing loss (CDS)). It consists of otologists, audiologists, pediatricians, clinical geneticists, neuroradiologists and, if indicated, neurologists or ophthalmologists.

2.2. Age

The age at detection was defined as the age at which the hearing loss was first diagnosed by the Audiology Center, either by ABR of PTA. Patients were categorized in 4 age groups: 0–1 year old, 1–6 years old, 6–12 years old and 12–18 years old.

2.3. Audiometric evaluations

The first test of the Dutch newborn screening protocol is performed at home and consists of transient evoked otoacoustic emissions (TEOAE). Children who fail the first test are retested using TEOAE, and subsequently using automated auditory brainstem measurements (AABR). Children admitted at a Neonatal Intensive Care Unit (NICU) are screened in the hospital by AABR. Children who fail all tests are referred within three weeks to an audiology center for further investigation for Automated Auditory Brainstem Response (AABR) in newborn children, or age appropriate assessment like Visual Reinforcement Audiometry (VRA), Behavioral Observation Audiometry (BOA) or pure tone audiometry (PTA) in older children. To determine the ABR thresholds (dB nHL) we used a clear appearance of wave V upon clicks. For the estimated behavioral hearing thresholds around 3 kHz (dB eHL) we use a correction of 10 dB. When PTA is performed, an average threshold at 500, 1000, 2000 and 4000 Hz is used for the analysis.

Children were diagnosed with SNHL if the sensorineural hearing threshold at the best hearing ear was 30 dB or more. Asymmetric bilateral SNHL was defined as 1 or more frequencies with a greater than 30 dB difference, 2 or more frequencies with a greater than 15 dB difference or 3 or more frequencies with a greater than 10 dB difference in threshold between the left and right ear. The hearing loss category was based on the hearing level of the most severely affected ear.

Hearing loss was categorized as a slight impairment (26–40 dB), moderate impairment (41–60 dB), severe impairment (61–80 dB) and profound impairment (81 dB or greater) according to the commonly used classification of the World Health Organization (WHO). In case of mixed type hearing loss, the inclusion and consecutive analyses were based on the sensorineural component only. Patients with pure conductive hearing loss were excluded from this study.

2.4. Evaluation of imaging

The decision to obtain imaging and the choice of imaging modality was individualized per patient and made by the CDS multidisciplinary team in close consultation with the parents.

The majority of the imaging was performed at the VUmc, Amsterdam, The Netherlands, following a standard temporal bone CT protocol, consisting of non-contrast axial 0.6 mm slices. Coronal and sagittal reconstructions (0.6 mm thickness) were performed, as well as axial reconstructions following the plane of the lateral semicircular canal. The MR imaging protocols of the brain and temporal bone include transversal T2 weighted, transversal FLAIR, sagittal 3D gradient-echo sequences and axial 3D constructive interference steady state (CISS) or FIESTA-C images centered at the level of the internal auditory canal. In some cases, imaging was performed in the referring center using a local CT and/or MR imaging protocol. These scans were re-evaluated by a radiologist and the otologist of the CDS multidisciplinary team.

The imaging was evaluated with a focus on abnormalities associated with sensorineural hearing loss at the level of the middle ear, the inner ear, the inner auditory canal (IAC), the cochlear nerve and the brain. The inner ear abnormalities included acquired pathologies and congenital malformations, which were classified as cochlear aplasia, cochlear hypoplasia, common cavity, incomplete partition type I and II, isolated enlarged vestibular aqueduct (EVA) (defined as a vestibular aqueduct diameter exceeding 1,5 mm, measured halfway between the common crus and the medial aspect of the opening of the operculum on the posterior wall of the temporal bone), isolated lateral semi-circular canal dysplasia, vestibular hypoplasia, and malformations of the cochlear nerve [12]. With regard to abnormalities at the level of the labyrinth and IAC each side was evaluated separately. The brain abnormalities were categorized in diagnostic findings (i.e. signs of CMV), associated findings (i.e. signs of asphyxia, vasculopathy, hydrocephalus, encephalitis) or non-associated findings (i.e. aspecific white matter abnormalities). The evaluation of the CT and/or MR imaging of the temporal bone and brain was performed by an experienced neuroradiologist and an otologist. In case of disagreement, joined evaluation by the neuroradiologist and otologist was performed in order reach consensus.

2.5. Statistical analysis

Statistical analyses were performed using SPSS 22.0. The criterion for statistical significance was set at $p < 0,05$. Descriptive analyses and cross tables were used to outline results of this study. In order to evaluate the correlation between age and hearing loss, logistic regression was used.