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Current Status and Problems in Hearing Examination for Young Children

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Abstract: Early detection of hearing loss is critical for a child's language development. With the worldwide spread of newborn hearing screening (NHS), significant progress in the prevalence of NHS has been made in Japan in recent years. As a result, early detection and intervention of hearing loss are being realized. However, because NHS coverage is not enough and children onset hearing loss after NHS exist, post-NHS young children's hearing screening is important. This review outlines the current status and challenges of hearing screening for young children (1-5 years old). The potential for new electrophysiological tests is also discussed.

Key Words: hearing loss, screening, electrophysiological measurement

Introduction

Hearing loss in childhood adversely affects speech and language development, impairing social and emotional development and the quality of life. Early detection of childhood hearing loss is therefore critical to enable appropriate treatment and intervention and reduce the impact of the disorder (1, 2). To provide early detection and intervention for infant hearing loss, newborn hearing screening (NHS) has been expanded in the US since the late 1980s. In 1995, the Forty-Eighth World Health Assembly urged member states to prepare national plans for early hearing loss detection within the primary healthcare framework (3). In response to this global trend, the NHS has expanded dramatically throughout Japan since the Ministry of Health, Labor, and Welfare (MHLW) implemented a pilot project in 2000 (4). In 2005, MHLW implemented the "Newborn Hearing Screening Project" as part of the "Comprehensive Support Project for Maternal and Child Health Care Measures," and in 2007, the project was incorporated into general revenue. In 2012, a section was added to the Maternal and Child Health Handbook to include the results of newborn hearing tests. According to the Japan Association of Obstetricians and Gynecologists, the percentage of facilities offering NHS increased from 32% in 2002 to 98.1% in 2020. In parallel with this, the screening rate of NHS increased from 13.0% in 2014 to 80.7% in 2019, as calculated by the total number of births per year and the number of newborns screened (4). The NHS data indicated that the incidence of hearing impairment in newborns in Japan was roughly 0.14% (4).

NHS testing methods are universal, including Auditory Brain Response (ABR) and/or Otoacoustic Emission (OAE). These tests are described in detail in the next section. Parents will be notified of the screening results as a "pass" or "refer". In addition to congenital hearing impairment, other causes of hearing loss include cerumen plugging and exudative otitis media. When a child is found to have "refer," a thorough examination by an otolaryngologist will be performed. As mentioned above, NHS in Japan has been rapidly expanded, and a system for early detection of hearing impairment has been established. However, there are still several problems. One is that although the NHS enforcement rate has increased, it is still far from the 95% rate recommended by the American Academy of Pediatrics, Joint Committee on Infant Hearing (JCIH)(1). Therefore, not a few patients could have been missed. Furthermore, it is well known that various factors are known to contribute to hearing loss in early childhood (Table 1),

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Table 1. Risk Factors for Early Childhood Hearing Loss (1).

1.	Family history *of early, progressive, or delayed onset permanent childhood hearing loss.
2.	Neonatal intensive care of more than 5 days.
3.	Hyperbilirubinemia with exchange transfusion regardless of length of stay.
4.	Aminoglycoside administration for more than 5 days.
5.	Asphyxia or Hypoxic Ischemic Encephalopathy.
6.	Extracorporeal membrane oxygenation (ECMO)*.
7.	In utero infections, such as herpes, rubella, syphilis, and toxoplasmosis. In utero infection with cytomegalovirus (CMV)*. Zika virus infection of mother and infant or mother only.
8.	Craniofacial malformations, congenital microcephaly, congenital or acquired hydrocephalus, and temporal bone abnormality.
9.	Syndromes with atypical hearing thresholds.
10.	Culture-positive infections associated with sensorineural hearing loss.
11.	Events associated with hearing loss such as significant head trauma or chemotherapy.
12.	Caregiver concern regarding hearing, speech, language, developmental delay and/or developmental regression.

* Infants at increased risk of delayed onset or progressive hearing loss.

and some congenital hearing loss is delayed in onset or progressive, like non-syndromic hearing loss by congenital cytomegalovirus infection (5). Because NHS is performed within the first month of life, such delayed or progressive hearing loss may be missed (JCIH recommended the 1-3-6 rule: hearing screening by one month of age, diagnosis of hearing loss by three months of age, and enrollment in early intervention by six months of age)(1). In line with this, the prevalence of hearing impairment is known to continuously increase during childhood, reaching about 0.2-0.3% in school-aged children (4, 6, 7). In order to provide appropriate treatment for patients with hearing impairment at a critical time in their language development (≤ 5 years old), early childhood hearing screening is essential. So, in Japan, hearing tests are conducted at the 1.5-year-old and 3-year-old child health checkups to detect hearing loss that develops in early childhood, and children suspected of having hearing loss are given a thorough hearing test. This article reviews the current status and issues of hearing detection for young children (1-5 years old).

Hearing detection procedures for young children

The audiologic assessment for young children has several components. If the child can understand language at 4-5 years of age, a pure tone audiogram and a test of middle ear function should be performed by an otolaryngologist to confirm the presence of otitis media effusion, which is the most frequent cause of childhood hearing loss (8). To measure middle ear function, tympanometry is commonly used (1, 9). Tympanometry is a test that measures the flexibility of the tympanic membrane while changing the air pressure in the ear canal. In tympanometry recording, the probe is placed in close contact with the ear canal, and the pressure in the ear canal is varied using a pump with a manometer while the sound is emitted from a speaker and its reflected sound is measured (Fig. 1A). In the normal ear, the movement of tympanic membrane is maximal when the pressure in the ear canal is near zero (Fig. 1B, type A). When the middle ear pressure is low due to dysfunction of the Eustachian tube, the tympanogram peaks when the ear canal pressure becomes negative (Fig. 1B, type C). When there is fluid retention in the middle ear (e.g., otitis media exudate) or adhesion of the tympanic membrane (e.g., otitis media adhesive), the tympanogram becomes flat (Fig. 1B, type B). A 226 Hz tone probe is the standard for tympanometry, but a 1000 Hz tone probe is recommended for infants under nine months (10).

If the child is under four years of age or cannot fully understand speech, objective hearing tests should be performed after an otolaryngologist examines the outer and middle ear: behavioral assessment of hearing, OAE, and electrophysiologic measurements (1). The following is a brief description of each test.

Behavioral assessment of hearing

Behavioral assessment of hearing is the gold standard for estimating the hearing threshold because the direct measurement of hearing is only assessed by evaluating the behavioral responses to sound (1). Different tests are used depending on the age of the child: conditioned orientation response audiometry (COR; also known as visual reinforcement audiometry, VRA; for infants 6 - 35 months) and condition play audiometry (CPA; for toddlers ≥ 36 months)(1, 9). In COR, children are first conditioned to show a head-turning response to the presentation of audible sounds, rewarded by a positive reinforcer (usually some visual display, Fig. 1C). Observing the conditioned responses, the hearing thresholds for the sound of different frequencies are evaluated. The CPA measures sensitivity to sound while using toys to attract the toddler's attention. The children are trained to respond in a game format (e.g., putting a block in a bucket) when the target sound is presented (Fig. 1D). The hearing threshold is then measured by presenting a variety of sounds and measuring responses

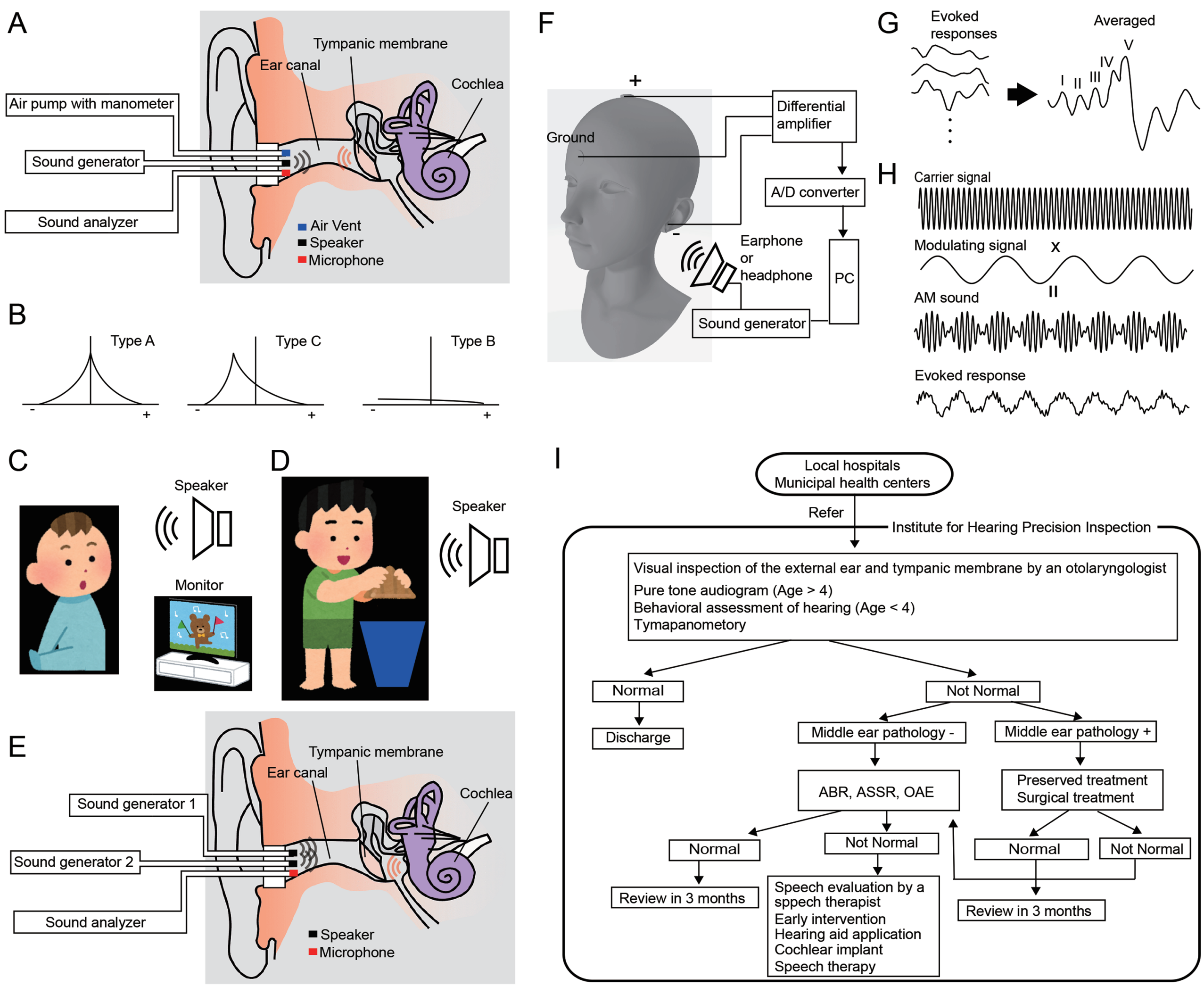

Figure 1. Hearing test for young children. (A) The recording scheme of tympanometry. The tympanometry measuring instrument has a pump with a manometer to change the air pressure in the ear canal, a sound generator with a speaker, and a sound analyzer with a microphone. During measurement, the probe is placed in close contact with the ear canal, and sound is applied to the ear canal while the pressure inside the canal is adjusted. The reflected sound is recorded with a microphone and analyzed. (B) Types of tympanogram. (C) Conditioned orientation response audiometry (COR). In COR, children are first conditioned to show a head-turning response to the sound by giving some rewardable visual display on the monitor when the sound is played. Once conditioning is complete, hearing thresholds are measured by varying the loudness of the stimulus sound. (D) Conditioned play audiometry (CPA). In CPA, children are trained to respond in a game format (e.g., putting a block in a bucket) when the target sound is presented. Once the children understand the rules, hearing thresholds are measured by varying the loudness of the stimulus sound. (E) The recording scheme of otoacoustic emission (OAE). The OAE measuring instrument has two sound generators with a speaker and a sound analyzer with a microphone. OAE is measured by using a microphone to record the sound generated by cochlear activity that occurs when sound enters the ear from a probe. (F) The recording scheme of electrophysiological measurements. Both auditory brainstem response (ABR) and auditory steady-state response (ASSR) are measured using the differential amplifier with the same electrode placement on the head. An active electrode is often placed at the vertex or midline frontal, a reference electrode at the earlobe or mastoid, and ground at the forehead. Stimulus sounds are controlled by a PC and given through earphones. Sound-evoked brain activity is amplified by a differential amplifier, digitized by a converter, and recorded on a PC. (G) The scheme of ABR recording. The ABR is obtained by averaging electrical brain responses to a click (0.5 ms) or a brief pure tone (3-5 ms). The ABR has I-V waves, each of which is thought to correspond to activity in a different part of the auditory neural pathway (I, auditory nerve; II, cochlear nucleus; III, superior olivary complex; IV, lateral lemniscus; V, inferior colliculus). (H) The scheme of ASSR recording. Amplitude-modulated (AM) sound probe is often used to record ASSR. AM sound is obtained by modulating the carrier signal with a lower frequency modulation signal. ASSR recordings measure the power of the modulation signal's frequency components in the electrical brain response. (I) Flowchart for diagnosis of hearing loss in children.

to them. Although these tests are important because they directly measure the extent to which a child can actually hear and respond to sound, hearing ability cannot be determined by these tests alone because the test results are highly dependent on the subject's condition at the time of the test. Further, it is known that about 40% of children with hearing impairment have physical, psychological, intellectual, or emotional needs or barriers that make the behavioral assessment challenging.

Otoacoustic emissions (OAE)

As an objective physiological screening measure, OAE is essential to identify children with hearing disorders. OAEs are sounds emanating from the cochlea when it is stimulated by a low-intensity click or pure tones, which is recorded by a microphone fitted in the ear canal (Fig. 1E)(11). They are generated by the motion of the outer hair cells in the cochlea in response to sound, so they are useful for evaluating the function of the inner ear. Commonly used OAE measurements are evoked otoacoustic emissions (EOAE), which measure the response to a single sound, and distortion product otoacoustic emissions (DPOAE), which measure the response to pure tones of different frequencies emitted from two speakers (Fig. 1E). OAEs are relatively quick (recording time = around 1 min) and easy to record and are used to screen for hearing loss and to cross-check behavioral observations. It is also used to monitor cochlear function during treatment with ototoxic drugs (e.g., aminoglycoside antibiotic therapy) that potentially cause damage to the outer hair cells. It should be noted, however, that disorders located more centrally than the cochlea, such as auditory neuropathy, cannot be detected by OAE.

Electrophysiological measurements

Electrophysiological measurements include ABR and auditory steady-state response (ASSR). These are the gold-standard tests for determining hearing thresholds (1). ABR reflects neural activity from the auditory nerve to the brainstem, while ASSR reflects activity from the brainstem to the auditory cortex. Both ABR and ASSR are recorded using the differential amplifier with the same electrode placement on the head (Fig. 1F). The ABR is obtained by averaging (1000-2000 times) the neural activity responses recorded using a click (0.5 ms) or pure tone burst (3-5 ms) as a probe (Fig. 1G). The ABR has I - V waves, with the wave V usually being the largest (Fig. 1G). Therefore, the hearing threshold is determined by examining the lowest sound pressure at which wave V appears in the ABR. Although ABR can determine auditory thresholds for sounds of different frequencies by using pure tone stimuli, it is known that sensitivity to low (<1 kHz) tones is low (12). Therefore, ABR primarily examines sensitivity to sounds between 2-8 kHz. Automatic ABR with click sounds is used by the NHS as a simple test to detect the presence of hearing loss in infants. ASSR is obtained by frequency analysis of brain activity recordings of amplitude-modulated (AM) tones, which can be used to examine auditory thresholds for tones of different frequencies by varying the carrier frequency of the modulated tone (Fig. 1H). The auditory nervous system is known to follow the modulation component of AM sound (13), and brain activity recorded from the scalp also shows activity synchronized with the modulation component (Fig. 1H). In contrast to ABR, ASSR recording uses a relatively long probe sound (tens of milliseconds to minutes) to measure brain activity. The ASSR test uses spectral analysis to detect activity synchronized with the AM component in the recording. The minimum sound pressure at which the AM component is detected in the recorded brain activity is determined as the hearing threshold for the carrier frequency. Compared to ABR, ASSR is known to have better sensitivity to low-frequency sounds (<1 kHz) (12), making it an indispensable test for audiometric testing in the low-frequency range.

The presence and degree of hearing loss in young children are diagnosed by comprehensively considering these tests. Among these, the ABR and ASSR are essential to confirm the hearing ability of referred patients. As a practical example, at Kanazawa Medical University, children (> 6 months) are observed for their turnaround response to sound (COR) in the outpatient clinic. Then, an appointment for ABR or ASSR testing is made for a thorough examination of the child's hearing. Tympanometry is not routinely performed in young children because they are often difficult to record. CT and MRI data are also used for diagnosis as needed. Figure 1I shows a flowchart of the diagnosis from these tests.

Problems in testing for hearing loss in young children

Testing by ABR and ASSR is essential to ultimately determine the presence or absence of hearing loss in young children. A drawback of ABR and ASSR, however, is that the test can be time-consuming (> 30 min) because it requires brain activity recordings of sufficient duration to obtain reliable results. The subject must remain at rest using sedative during the test to prevent electrical noise contamination, which can make testing young children difficult. They often cannot remain in the same position for long periods of time and require sedation for ABR and ASSR in many cases (1). For ABR and ASSR on sedated young children, it is necessary for the otolaryngologist, pediatrician, and co-medical staff to work together. This requires the coordination of many personnel schedules, making frequent testing difficult. In addition, when sedation is necessary, parents are often reluctant to have the examination because of the risks that occur during sedation. For this reason, some children with speech delays are not tested when severe hearing loss is not suspected. The following section discusses the possibility and utility of developing electrophysiological tests that can be performed more conveniently.

Challenges in recording electrical neural activity from awake young children

If ABR and ASSR measures on toddlers that do not require sedation were developed, hearing testing for young children would be dramatically enhanced. There are several issues that need to be resolved in order to get these records from an awake young child. First, it is necessary to reduce noise caused by body movement in order to obtain sufficient quality recordings for diagnosis. Body movement introduces noise into the recording due to muscle movement and movement of the electrode wires. A known method to reduce such noise is to place a recording amplifier on the head, which digitizes the recorded potential. This method is commonly used in electrophysiological experiments on animals in action and has been shown to dramatically reduce the noise generated in the wiring of the recording device (14). Currently, inexpensive digital amplifier chips for bioelectric recording are available (e.g., https://intantech.com/), and the development of such a device can be done at a low cost. Several research groups are developing wireless ABR recording devices, although they have not yet reached practical application. Our group is also currently developing a helmet-integrated recording device that combines a small digital amplifier with a microcomputer and wireless communication (Fig. 2).

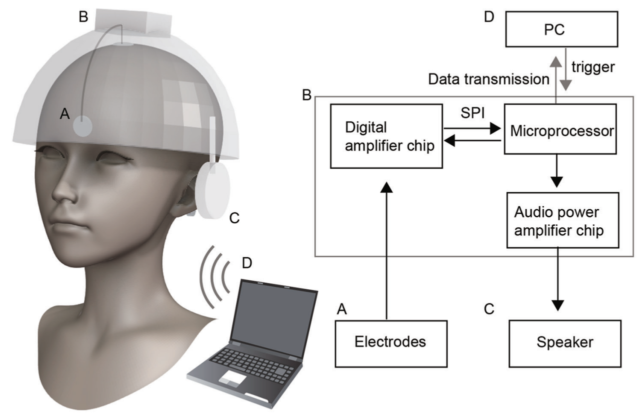

Figure 2. Scheme for a prototype Wireless ABR/ASSR measurement system. An integrated recording device with electrodes (A), a control box (B), and speakers (C) mounted on a helmet is remotely controlled by a PC (D). A programmable microprocessor is built into the control box. When the recording starts, the microprocessor generates sound waves, which are delivered to speakers through an audio power amplifier chip and produce sound stimulus. Recordings of the electrical brain activity are obtained through the electrodes on the head. The electrical signals are amplified and digitized by a digital amplifier chip. The microprocessor exchanges data with the digital amplifier chip via Serial Peripheral Interface (SPI) communication. The microprocessor is remotely controlled from the PC to start recording and send recorded digitalized data to the PC.

Second, there is the psychological impact of the stimulus sound on young children. ABRs and ASSRs typically measure auditory thresholds by beginning by recording responses to 80 dB sounds in order to obtain reliable responses that can be used as the basis for analysis. These tests require repeated recordings of responses to the probe sounds in order to obtain analyzable data. A sound of 80 dB corresponds to a noisy level of sound pressure (e.g., inside a subway car, garbage disposal machine, etc.), and continuously exposing a young child to such a sound is likely to cause mental distress. In addition, as mentioned above, at least 30 minutes of recording time is required to determine the hearing thresholds for multiple-frequency sounds. Continuously playing a monotonous sequence of tones for such a long period of time is also likely to be distressing to young children. To avoid these problems, one solution is to make the presentation discontinuous when playing a sound with high sound pressure. Previous psychological research has shown that sound "annoyance" is largely related to the duration of the sound and the inter-stimulus interval (15) and that the shorter the duration of the sound and the longer the interval, the less "loud" the sound is. In addition, it was reported that startle responses to loud noises can increase annoyance (16). In light of these findings, rather than repeatedly playing a series of sounds of the same sound pressure level as in the conventional method, repeatedly playing sound sequences ordered from lowest sound pressure to highest sound pressure level may be more effective in reducing auditory discomfort because it avoids abrupt changes in sound pressure and increases the interval between the presentation of sounds with high sound pressure level. Further, new methods using synthetic language sounds are being attempted to be developed for ASSR testing (17). Since the ASSR test examines responses to AM sounds, it is possible to detect and analyze brain activity responses to AM sound components in synthetic language sounds. If such a test using synthetic speech sounds becomes possible, the test can be performed while a story is read to the patient, which is expected to reduce stress during the test dramatically. It is important to note, however, that since the ASSR is known to be dependent on the subject's attention (18), it is possible that language sounds may affect the subject's attention and alter the ASSR, so it is essential that the development of such a new method be thoroughly compared with conventional methods.

Conclusion

The detection of hearing loss in children has progressed rapidly in recent years due to the significant expansion of the NHS. However, post-NHS hearing screening of young children is important because of omission and oversights in the NHS and progressive hearing loss disease. However, ABR and ASSR for definitive diagnosis have not been performed in sufficient numbers due to technical limitations. The development of stable recording systems in the awake state and stress-free sound presentation methods is expected to enhance the testing of hearing loss in young children.

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Disclosure of Conflicts of Interest

The authors declare no conflicts of interest.

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