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Auditory brainstem response record of Down syndrome under an anesthesiologist's control

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We recorded auditory brainstem response under an anesthesiologist's control for a child with Down syndrome for whom we could not induce sleep by the usual methods. The patient was a 5-year-old girl. After administering midazolam and propofol, we recorded the patient's auditory brainstem response (ABR) while she was asleep and maintained spontaneous respiration. The results were that the threshold of the right ear was 30 dBnHL, and the left ear was 40 dBnHL. These results indicated from a normal to mild hearing loss. Therefore, we thought that there was no indication of a need for a hearing aid at present. The advantages of an ABR recorded under an anesthesiologist's control are that we can make the examinee rest and induce sleep, and that there are no influences of the rest and sleep on the ABR. Moreover, with anesthesiologists present, they can deal with respiratory failure if it occurs. On the other hand, the disadvantages are that patients have to be admitted to a hospital therefore increasing the economical burden for the patients or family. Furthermore, this requires anesthesiologists to work in different circumstances such as in hearing testing rooms.

Key words: auditory brainstem response, anesthesiologist, Down syndrome

Introduction

Examinees most in need of objective hearing tests are infants, hyperactive children, and children such as those who are emotionally disturbed, mentally retarded, and doubly handicapped.[1] Objective hearing tests for infants that can be examined at a young age are desirable for hearing evaluations as soon as possible.

Auditory brainstem response (ABR) is considered to be the best index of objective hearing evaluations at present from the following: influences of depth of sleep on the response wave form and threshold are negligible, and the response threshold is comparatively lower than other auditory electrical responses.

A sleep condition is more suitable than an awake condition for these evaluations, because clear responses can be recorded without the interference of electromyogram and body movement. Therefore, for children, we always record under induced sleep by giving them triclofos sodium syrup. However, there are often cases in which inducing sleep is difficult. In the present case, ABR was recorded under an anesthesiologist's control for a child with Down syndrome for whom we could not induce sleep by the usual methods.

Case

Patient: Down syndrome, 5-year-old girl.
Chief complaint: retarded speech development.
History of present illness: Parents have been thinking that her speech development is slow. In January, the parents took their daughter to an ear, nose and throat clinic following the advice of the child's kindergarten teacher. There they were referred to Kitasato University Hospital for a more thorough examination and first came to our hospital in February. They told us that their daughter often went up close to the television and sang and danced with it at home.
Birth: gestational age, 40 weeks; Caesarean section; birth weight, 2,026 g.
Motor development: head control, 9-12 months old; sitting position, 12 months old; erect position, about

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12 months old; beginning to walk, 24 months old.

Speech development: beginning to talk, 24 months old.

Past medical history and Family history: None.

Otolaryngological findings at first visit: There were no abnormal findings except that bilateral external auditory canals were slightly narrow. We tried conditioned orientation reflex audiometry but did not succeed, because the child was restless and would not sit still in the chair and stood up and made a lot of noise.

Progress: We tried to record the ABR by giving the child triclofos sodium syrup 15 ml per 13 kg body weight for a total dose of 100 mg. But we could not induce sleep or do the recording. Because an ABR record is necessary for a hearing evaluation, we planned to record the ABR under an anesthesiologist's control and performed the usual preoperative examinations (hematological examination, urinalysis, chest x-ray and electrocardiography). The results were all normal. She was admitted to our hospital the day before the ABR was recorded, which was done in a hearing test room, not a soundproof room. After anesthesiologists began giving O_2 at 1.5 l/min, they injected midazolam intravenously (total 1.25 mg). After 10 minutes, they administered propofol by an intravenous injection and a continuous intravenous infusion (3 ml/h) for a total of 100 mg. After 10 minutes of sleeping with spontaneous respiration, we attached the electrodes, held the headphone, and started recording. After 40 minutes of sleep, we finished the examination. The anesthesiologists discontinued the propofol just before we finished, and the patient woke up 5 minutes later (Figure 1).

Anesthesiologist's control

O_2 transnasal cannula 1.5 L/min (total 180 L)
Midazolam IV (total 1.25 mg)
↓ after 10 minutes
Propofol IV and continuous IV (total 100 mg)
↓ after 10 minutes
We started recording during sleep.
↓ after 40 minutes
We finished recording.
↓ after 5 minutes
The patient awoke.

Figure 1. The ABR record under an anesthesiologist's control

We brought the same anesthesia machine used in the operating room into the hearing test room that was out of a soundproof room where the recording was made. The procedure took about 80 minutes.

Condition of ABR record: We attached the electrodes as in the positive upper method as is usually done: active electrode at the vertex, reference electrode on the mastoid, and a grounding electrode on the forehead. We used the Neuropack MEB-2208 (Nihon Koden Co., Tokyo) for the record. The stimulation sound was clicking at a frequency of 16.3 Hz, the additional

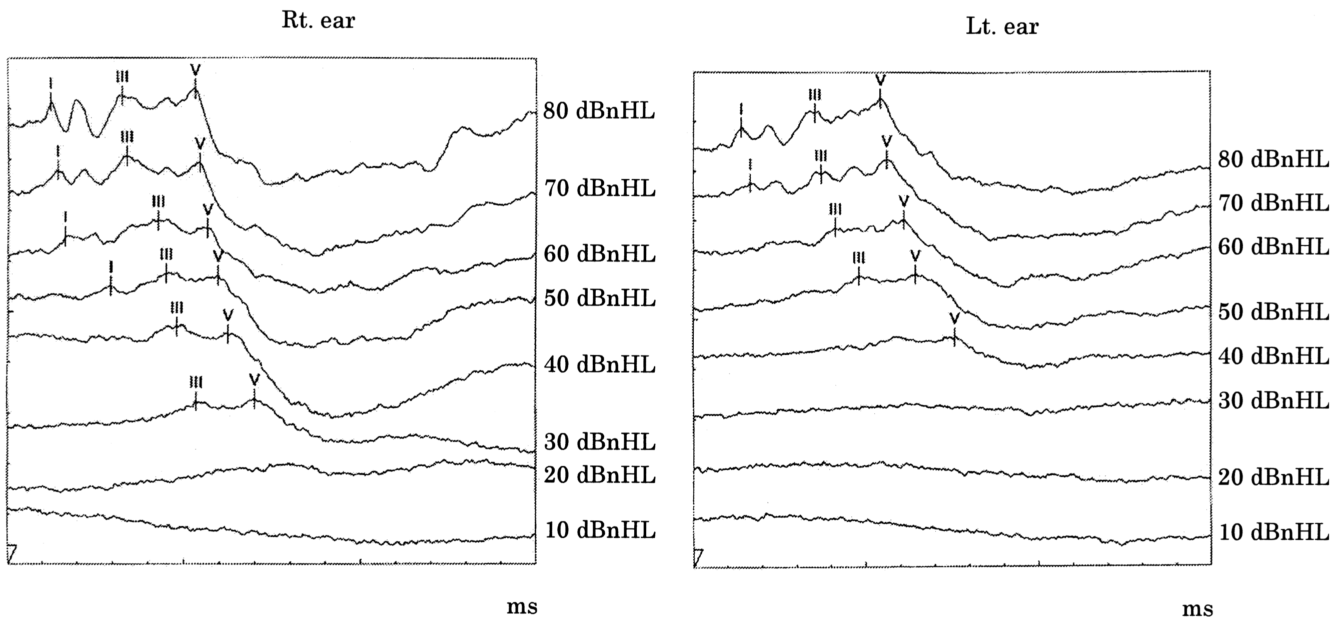

Figure 2. The results of the ABR recording

The threshold of the right ear was 30 dBnHL and the left ear was 40 dBnHL. Bilateral latency of waves I and V was shortened. The horizontal axis shows that 1 unit on the scale is 1 millisecond (ms).

number of times was 2,000, and the stimulation sound pressure ranged from 80 dBnHL to 10 dBnHL and 10 dBnHL step, the analyzed times were 15 ms, and the filters were from 50 to 3,000 Hz.

Results

The threshold of the right ear (wave V) was 30 dBnHL, and the left ear was 40 dBnHL (Figure 2). These results indicated from a normal to mild hearing loss, so we judged that there was no indication of requiring a hearing aid at the present time. Because there are possibilities of fluctuations, we are continuing the follow-up.

Incidentally, the latencies of the right ear were: wave I, 1.23 ms; wave V, 5.32 ms; and the interpeaks of waves I-V were 4.09 ms at 80 dBnHL (50 dBSL), and the left ear were 1.42 ms, 5.45 ms, and 4.03 ms, so the bilateral latencies of waves I and V were shortened.

Discussion

The major advantage of an ABR record is an electrophysiological reaction from the auditory system. Another advantage is that we can get information about thresholds, while the greatest disadvantage is that it takes a relatively long time to do the test. It takes a long time to attach the electrodes correctly. So only a little time is saved even if the number of times and stimulation sound pressure decreases. The examinees often wake up and we have to wait for them to fall asleep again. Practically we could not record in this case because she could not understand that she must keep still, and we could not make her sleep by triclofos sodium syrup. Another disadvantage is that click sounds are used to test auditory levels of high tones, so we could not test some of the low-tone frequencies. We can solve this problem, to a certain extent, if we use tone-pips for the stimulation sounds. But we can not acess the audiogram exactly in the case of the dip type.[2] Regarding this problem, we can presume the auditory level of each frequency in all auditory types when we find the amplitude-modulation following the response threshold with a changing constant frequency in the auditory steady-state response (ASSR). Even though the difference between the reaction threshold and the auditory level is almost the same as that in the ABR, it must be checked carefully because Rance et al.[3] reported that the less hearing loss there is, the larger the difference there is between the auditory level and the ASSR threshold. However, ASSR had not yet been used in our hospital when we made the record under an anesthesiologist's control. The last disadvantage is that there are some cases in which severe encephalopathic children, and in cases of various degeneration diseases, that patients present as being clinically normal, even if there is no audiological response.[4,5]

The frequency of Down syndrome is about 1/1,000, the features of which are: a moderate failure to thrive, moderate mental retardation, upturned palpebral fissure, flat root of nose, dorsum of nose, and auricular deformity, among others. Although there are various ranges of hearing loss, about 50% of patients present with some degree of hearing loss, and most of them contract otitis media with effusion.[6] But there are more than a few cases in which deformity of the inner ear probably caused severe sensorineural hearing loss.[7]

Even though the ABR wave forms in Down syndrome patients have various patterns: normal, no response, and so on, depending on the case, the following abnormal findings have been reported.[7~11]

1. No response: Many may have severe hearing loss.
2. Prolongation of latency of wave I, slight elevation of the reaction threshold: This may be caused by otitis media with effusion and deformity of the external and middle ear.
3. Shortening of latency of waves I, V, and the interpeak of waves I-V: These are very specific abnormal findings as in the present case. There was only one case of the 13q syndrome in other diseases that the latency of each wave was shortening.[12] Kaga et al.[10] reported that the frequency of shortening of latency of waves I, V, and the interpeak of waves I-V were 2/37 (5%), 7/37 (19%), and 8/37 (22%), respectively. Though the mechanism of shortening of latency remains unknown, there is one report in which the length of the cochlea was shortened in Down syndrome.[13] Deformity of the auditory tract may contribute to shortening of latency.
4. Time series changes in the ABR wave form: Even though we find these in other cases of chromosome aberrations, there are some cases in which the ABR wave forms normalize in progress.[7,8] Though most of the cases may be caused by recovery from serous otitis media and/or the function of the conductive system, the center level, like the development of myelination, may be a contributing factor.

Even though the studies cited did not give details about sedation, Nakagawa et al.[14] reported that they studied the influence of propofol on the ABR before performing a neurosurgical operation, and there was little influence of 3 μg/ml propofol. The amount of propofol we used was estimated to be roughly less than 3 μg/ml.

The advantages of performing the ABR record under an anesthesiologist's control are that we can make the patients rest and induce sleep, there are no influences of the rest and sleep on the ABR, and anesthesiologists are available to deal with respiratory failure if it occurs. On the other hand, the disadvantages are that patients have to be admitted to a hospital, which increases their economical burden, and it forces anesthesiologists to work in different circumstances than they do usually, i.e., in operation rooms.

Conclusion

We recorded the ABR under an anesthesiologist's control because we could not induce sleep in the patient with Down syndrome by the usual methods. This method, under general anesthesia, has the advantages that we can make the patient sleep, and there are no influences of that sleep on the ABR. It takes longer to test ABR and ASSR simultaneously. Therefore, this may be a useful method to diagnose infants, patients presenting with hyperkinesia, and handicapped children such as emotionally disturbed children and mentally retarded children for whom we can not induce sleep by the usual methods.

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