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Research Article

Air-And Bone-Conduction Auditory Brainstem Response in Children with Congenital External Auditory Canal Atresia

Abstract

Objective: This study aimed to determine the clinical value of air- and bone-conduction auditory brainstem responses (ABRs) in children with congenital external auditory canal atresia (EACA).

Methods: Air- and bone-conduction click-evoked ABRs in 38 children having congenital EACA were compared with 34 children having normal hearing.

Results: ABR threshold for air and bone conduction were 66.53 ± 7.12 and 12.55 ± 6.96 dBnHL, respectively, in children with congenital EACA, as well as 25.32 ± 2.66 and 10.71 ± 4.51 dBnHL, respectively, in children with normal hearing. The two groups showed statistical difference in air-conduction ABR thresholds. Meanwhile, air-bone ABR threshold gap was greater in children with EACA than in children with normal hearing, and bone-conduction ABR wave latencies did not statistically differ between the two groups.

Conclusion: Bone-conduction ABR is valuable in assessing the function of cochlea, auditory nerve, and brainstem in individuals with congenital EACA. The study has important clinical value in the objective differential diagnosis of conductive deafness with combined application of air- and bone-conduction ABRs.

Introduction

External auditory canal atresia (EACA) is a birth defect characterized by hypoplasia of the external auditory canal, often associated with dysmorphic features of the auricle, middle ear, and (occasionally) inner ear structures [1]. EACA is a common birth defect that occurs in approximately 1 in 10 000 to 20 000 births. Most cases are sporadic and of unknown etiology. Nevertheless, several of these cases are syndromes, such as Goldenhar, Treacher-Collins, and Branchio-Oto-Renal syndromes. Unilateral EACA is more common than bilateral cases with an approximate ratio of 3 to 4:1. For unknown reasons, right ears are more commonly affected than left ears and boys are more commonly affected than girls are [2].

Diagnosis of a congenital ear malformation is usually made at birth when a malformed pinna or atretic canal is noticed during the secondary survey of the newborn. Some cases may not be noticeable at birth, such as those with a normal pinna and a blunted or partially patent canal. Several of these patients are detected during health screening examinations in schools. Air-conduction auditory brainstem responses (ABRs) are widely accepted in clinical audiology; however, it cannot distinguish conductive and sensorineural hearing loss in young children. In particular, testing infants is difficult with conventional behavioral audiometric techniques to evaluate the location and type of hearing loss; hence, bone-conduction ABR may contribute additional clinically valuable information. This method, however, has been ignored for a long time [3,4]. The purpose of this paper is to investigate the role of bone-conduction ABR in EACA infants.

Subjects and Methods

Subjects

Data from 38 infants (50 ears) aged 1–12 months (mean age = 4.6 months) diagnosed with EACA were collected from Children's Hospital of Fudan University between August 2014 to March 2015. The subjects were 28 boys, of which 12 cases were bilateral. Furthermore, the control group consisting of 34 normal infants (68 ears) aged 1–9 months (mean age = 4.1 months) were subjected to otoscopy, tympanometry, otoacoustic emission, and ABR; No hearing losses were reported among the children. For 38 EACA infants, CT scan and ABR were used to diagnosis and assessment. This work was approved by the Ethics Committee of Children's Hospital of Fudan University and conducted in accordance with the ICH guidelines for Good Clinical Practice and the Declaration of Helsinki. All of the subjects in the study signed an informed consent form.

Air- and bone-conduction ABR measurement

Air- and bone-conduction ABRs were performed in a soundproof room. All test subjects orally received 0.5 ml/kg dose of 10% chloral hydrate to induce a 'deep' sleep. Air- and bone-conduction ABRs were measured with a GSI Audera Brainstem Analyzer using Model TDH-39P headphone and AD229 earphone, respectively. Silver disc electrodes were placed on the forehead (active), nasion (ground), and earlobe (reference) for air-conduction ABR. When bone-conduction was performed, the AD299 earphone was placed on the mastoid bone without moving the reference electrode. The band pass filter settings were 100 and 2500 Hz (bone ABR, 50–1500 Hz) with a 10 Ms window

(bone ABR, 20 ms). Average response was obtained twice by recording the responses to two series of 2006 clicks. For air-conduction ABR, alternating polarity clicks at 75, 65, 55, and 45 dBnHL were used. Defined as the lowest stimulus level at which peak wave V could be visualized, threshold was recorded. For bone-conduction ABR, alternating polarity clicks at 45, 35 and 25 dBnHL were used, and the threshold was recorded as well. Contralateral acoustic stimulus was not performed on infants with bilateral EACA because of its difficulty. In lieu of contralateral acoustic stimulus, white noise under 20 dB was used for unilateral infants.

Statistical analysis

The results were exhibited as means ± standard derivation (SD). T-test was used to evaluate the significance of inter-group differences. Values with P<0.05 were considered statistically significant.

Results

A total of 38 EACA infants (50 ears) and 34 normal infants (68 ears) were tested with air- and bone-conduction ABR, and thresholds were recorded. In air-conduction ABR, the mean threshold of EACA infants significantly increased compared with that of normal infants (P<0.001). However, no statistical differences were observed between two groups (P=0.10) in bone-conduction ABR. Furthermore, threshold differences between air-conduction and bone-conduction ABRs were calculated, and the results indicated that the threshold differences of EACA infants were much higher than of normal infants (P<0.001) as shown in Table 1).

Second, the waveforms I, III, and V of bone-conduction ABR under 45, 35, and 25 dBnHL were recorded in all cases. With increased stimulus intensity, the amplitude became larger and the latency became longer, and wave V gave a larger amplitude than the other waves. Moreover, the latency functions of waves I, III, and V, and inter-peak latency of I-III, III-V and I-V evoked by bone-conducted stimulus were compared between EACA and normal infants. No significant differences were observed (P>0.05) as shown in Tables 2,3.

Moreover, the waveforms I, III and V of air-conduction ABR were recorded in EACA and normal infants as well, significant difference was observed in waves I, III and V latency between two groups, but no significant difference was observed in inter-peak latencies of I-III, III-V and I-V, the results were shown in Tables 4, 5.

Discussion

EACA is a common birth defect in otolaryngology where in the external auditory canal or the structures of the middle or inner ear fail to develop completely. The development may be arrested at any point in the process; therefore, clinicians may encounter patients with various degrees of severity of this defect. The incidence of EACA is 1 in 10 000–20 000 live births worldwide. The highest incidence

Table 2: Latency of wave I, III, and V evoked by bone-conduction ABR.

Group	No.	I mean (ms) ± SD	III mean (ms) ± SD	V mean (ms) ± SD
ECAA	66	2.27 ± 0.25	4.99 ± 0.44	6.78 ± 0.38
normal	80	2.19 ± 0.45	4.83 ± 0.51	6.65 ± 0.33
P		0.19	0.38	0.28

Table 3: inter-peak latencies of I-III, III-V and I-V evoked by bone-conduction ABR.

Group	No.	I—III inter-peak latency mean (ms) ± SD	III—V inter-peak latency mean (ms) ± SD	I—V inter-peak latency mean (ms) ± SD
ECAA	66	2.71 ± 0.21	1.81 ± 0.18	4.54 ± 0.22
normal	80	2.67 ± 0.15	1.82 ± 0.22	4.48 ± 0.17
P		0.51	0.78	0.42

Table 4: Latency of wave I, III, and V evoked by air-conduction ABR.

Group	No.	I mean (ms) ± SD	III mean (ms) ± SD	V mean (ms) ± SD
ECAA	66	2.25 ± 0.27	4.89 ± 0.29	6.85 ± 0.33
normal	80	1.58 ± 0.15	4.20 ± 0.24	6.35 ± 0.28
P		P<0.001	P<0.05	P<0.05

Table 5: inter-peak latencies of I-III, III-V and I-V evoked by air-conduction ABR.

Group	No.	I—III inter-peak latency mean (ms) ± SD	III—V inter-peak latency mean (ms) ± SD	I—V inter-peak latency mean (ms) ± SD
ECAA	66	2.60 ± 0.16	2.01 ± 0.24	4.56 ± 0.18
normal	80	2.55 ± 0.18	2.08 ± 0.22	4.53 ± 0.14
P		P>0.05	P>0.05	P>0.05

rate is in Japan and India with 1 in 1200–4000 patients [1]. EACA commonly coexists with microtia. Thus, the appearance is affected. Moreover, EACA can cause conductive hearing loss, which results in profound implications for development of speech and language in infants and young children, particularly in bilateral cases. Surgery or ossiphone is usually used to treat EACA infants after a series of necessary detailed evaluations, including audio logical evaluation [5].

ABR is an auditory evoked potential extracted from ongoing electrical activity in the brain and recorded via electrodes placed on the scalp. ABR is a widely used parameter in newborn hearing screening, auditory threshold estimation, determining hearing loss type and degree, and auditory nerve and brainstem lesion detection. Air-conduction ABRs are widely used clinically; however, these ABRs cannot distinguish types of hearing loss. When pure tone audiometry cannot be tested either, the situation is even worse [6]. Bone-conduction ABRs are similar to air-conduction ABRs, although the former transmits signal rather than the latter [7].

In our study, we testified that the threshold of air-conduction ABRs in EACA infants were significantly higher than in normal infants. In bone-conduction ABRs, however, no statistical differences were observed between the two groups. Besides, all the cases were examined by temporal bone CT scan and all infants were diagnosed

Table 1: Thresholds and threshold differences in EACA and normal infants.

Group	No.	Air-conduction ABR mean threshold (dBnHL) ± SD	Bone-conduction ABR mean threshold (dBnHL) ± SD	threshold difference
EACA	66	66.53 ± 7.12	12.55 ± 6.96	53.88 ± 8.64
normal	80	25.32 ± 2.66	10.71 ± 4.51	14.27 ± 4.20
P		<0.001	0.10	<0.001

with external auditory meatus atresia with or without middle ear malformation. Furthermore, no inner ear malformations were found by CT. The results of our study indicated that the detection of bone-conduction ABRs was consistent with CT scans. However, the threshold differences of EACA infants were much higher than in normal infants. This result indicated that all cases had conductive deafness.

Latency largely reflects activity of the cochlea, auditory nerve, and brainstem and is subjected to cochlear conduction time, conduction velocity in nerve fibers, synaptic delay between hair cells and auditory nerve fibers, and synaptic delay between nerve fibers in the brainstem pathway [8]. In our study, the latency function of wave I, III, and V evoked by bone-conducted stimulus is compared between EACA and normal infants. No significant differences were observed. This result further confirmed that bone-conduction ABRs could reflect inner ear function without being affected by the outer and middle ear.

Although bone-conduction ABRs are derived from air-conduction ABRs, these two ABRs have many differences, including the aspect of relevance ratio, response threshold, and latency. Debates on bone-conduction ABR are still inconclusive and bone-conduction ABR reportedly has lower relevance ratio, higher response threshold, and longer latency compared with air-conduction ABR [9]. Nevertheless, the waves evoked by bone-conduction ABR in our study were clear, stable, with good repeatability, and well differentiated. The threshold of bone-conduction ABR was lower than in the air-conduction ABR although their latencies of wave V were similar. Infants and adults have physiological differences, such as skin thickness, body fat, mastoid air cells, bone density, and brain development. Consequently, their responses to bone-conduction ABR are dissimilar. Similar with the present study, Yang et al. [10], found no prolonged latencies of wave V in infants in their survey.

Air-conduction ABR is widely used in infants with EACA or those infants with middle ear malformation; however, it cannot distinguish the types of hearing loss. One problem is that infants are difficult to test with conventional behavioral audiometric techniques; hence, bone-conduction ABR may contribute additional clinically valuable information. Vibration of the skull results in auditory sensation and a way to bypass the outer and middle ears to stimulate the cochlea. The goal of bone ABR is to estimate cochlear function and to help identify the type of hearing loss present [11,12]. Moreover, bone-conduction ABR can be used to evaluate the hearing level after wearing bone-anchored hearing aid, particularly in children. Detection of conductive hearing loss is easier below 1000 Hz, but the chirp-stimuli ABR mainly reflects the mean threshold around 1000–4000 Hz. The difference can result in underestimation of low-frequency hearing loss, especially in patients with tympanitis or otosclerosis. For some moderate or severe middle ear hearing loss patients, such as EACA, bone-conduction ABR can estimate cochlear function well.

Bone-conduction ABR has several clinical limitations, such as 1) the maximum output for bone is around 50 dBnHL and distinguishing between sensor neural or mixed hearing losses is difficult when the losses by bone exceed 45 dBnHL; 2) another problem is masking dilemmas as the energy can cross from non-test ear to test ear; 3)

same pressure of bone vibrator and head cannot be assured, especially when the test objective is infants or children [13]. Apart from these limitations, the present study is limited to infants; thus, our parameters can only represent the features of EACA infants without inner ear malformation. Further research on bone-conduction ABR in children of other ages is needed.

In conclusion, this study demonstrated that bone-conduction ABR can estimate cochlear function and identify the type of hearing loss. The study has important clinical value in the objective differential diagnosis of conductive deafness with combined application of air- and bone-conduction ABRs.

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