Paediatrica Indonesiana

VOLUME 43 January - February • 2003 NUMBER 1-2

Original Article

Brainstem auditory evoked potentials in children with microcephaly

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ABSTRACT

Background Hearing loss (HL) is commonly found in children with microcephaly. The aim of this study was to reveal hearing loss and auditory brainstem pathways disorders in children with microcephaly and other handicaps.

Methods There were 194 children who were referred for hearing evaluation. Subjects with history of congenital perinatal infection (TORCH) were excluded. Data were collected from the results of Brainstem Auditory Evoked Potentials (BAEP) recordings, including sex, age, clinical manifestations, latency and interlatency between waves I, III, V, and the hearing levels of each ear.

Results Moderate to profound HL were found in fourteen ears (58%) of patients with microcephaly. Moderate to profound HL (28%) and endocochlear damage (15%) were found in the ears of patients with microcephaly and delayed speech. Moderate to profound HL (39%) and endocochlear damage (11%) were detected in the ears of patients with microcephaly and delayed development. Moderate to profound HL (21%) and endocochlear damage (16%) were found in the ears of microcephalic patients with both delayed speech and delayed development. Moderate to profound HL (26%) and endocochlear damage (32%) were detected in the ears of patients with microcephaly and cerebral palsy.

Conclusion This study revealed the importance of early HL detection in microcephalic patients especially those with other handicaps such as delayed speech, delayed development, and cerebral palsy **[Paediatr Indones 2003;43:28-30].**

Keywords: *hearing loss, microcephaly, auditory brainstem pathways.*

The goal of universal hearing screening is to identify children with hearing loss by 3 months of age and to provide appropriate intervention by 6 months of age. Intervention begins before 6 months of age, substantially preceding the speed and language skills of children.[1] Brainstem Auditory Evoked Potentials (BAEPs) are used in pediatrics to detect and measure hearing loss in children, especially for those under the age that can be tested behaviorally, and also to evaluate the auditory brainstem pathways in children who may have neurologic problem.[2]

The term microcephaly denotes an occipitofronto circumference that falls two or more standards deviations below the mean head circumference of a certain individual age and gender. Microcephaly necessitates a small brain and implies neurologic impairment.[3] The aim of this study was to reveal hearing loss and auditory brainstem pathways disorders in children with microcephaly and other handicaps

Methods

The study was done in the Department of Child Health of Cipto Mangunkusumo Hospital Jakarta. During January 1997 to December 2001, there were 194 children who were referred for hearing evaluation. Subjects with history of congenital perinatal infection (TORCH) were excluded. Data

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were collected from the results of BAEP recordings, including sex, age, clinical manifestations, latency and interlatency between waves I, III, V, and the hearing levels of each ear.

The subjects were premedicated, using oral cloralhydrate 50-60 mg/kg, with intramuscular chlorpromazine 1 mg/kg, or the latter alone. Brainstem Auditory Evoked Potentials were recorded with Nihon Koden Neuropack Four Mini. Electrodes were placed on the vertex and ear electrodes were placed on the lateral surfaces of the left and right earlobe (A1, A2). The stimulus click rate was 20/second with a 3dB filter and intensities of 80 dB normal hearing level (nHL). System band-pass for recording was 30-3000 Hz, with an average stimulus of 2040 clicks. Every recording was done at least 2 times for each ear,[4] and recorded one by one ear for every child. The recordings were analyzed for each ear: the measurement of latency and intervals for waves I, III, and V at 90 dBHL (or 105 dBHL).

Classification of BAEPs[5]: (1) normal BAEPs: at 90 dBHL, wave I, III, and V peaks latencies and intervals are normal for age within 2 SD range. Electrophysiological hearing threshold is normal with wave V found at 30 dBHL; (2) transmission impairment: at 90 dBHL, wave I, III, and V latencies are more than 2 SD above normal of age values; I-III, III-V and I-V intervals are normal for age; electrophysiological hearing threshold is high; wave V is absent at 30 dBHL, but recordable at some higher intensities, (3) endocochlear damage: there is no reproducible wave, not even wave I, at 90 or 105dBHL. Partial damage is characterized by wave I, III, and V latencies and intervals which are normal at 90 or 100 dBHL, but electrophysiological hearing threshold above 30 dBHL. (4) retrocochlear impairment: wave I is present, but wave III and/or V latency is long for age as is the I-V interval. The electrophysiological hearing threshold is variable.

Severity of hearing loss (HL) can be classified as [6]: (1) none: 0 - 20 dB, (2) mild: 25 - 40 dB, (3) moderate: 45 - 60 dB, (4) severe: 65 - 80 dB, and (5) profound : > 80dB.

Results

In the study period, there were 194 patients (108 males and 86 females) referred to have hearing impairment evaluation. They were between 2 and half months and 7 years old.

Moderate to profound HL were found in fourteen ears of patients with microcephaly. Moderate to profound HL (28%) and endocochlear damage (15%) were found in the ears of patients with microcephaly and delayed speech. Moderate to profound HL (39%) and endocochlear damage (11%) were detected in the ears of patients with microcephaly and delayed development. Moderate to profound HL (21%) and endocochlear damage (16%) were found in the ears of microcephalic patients with both delayed speech and delayed development. Moderate to profound HL (26%) and endocochlear damage (32%) were detected in the ears of patients with microcephaly and cerebral palsy (**Table 1**).

One hundred and twenty six patients (65%) were within the age range of 1 to 4 years. Moderate to profound HL (41%) and endocochlear damage (19%) were detected in patients less than 1 year old. Moderate to profound HL (44%) and endocochlear damage (28%) were detected in patients aged 1 to 4 years old.(**Table 2**).

Tabel 1. Distribution of hearing loss based on the patients diagnosis

Diagnosis	<40dB (normal-mild)	(%)	41-105dB (moderate-profound)	(%)	105dB(-) (endocochlear damage)	(%)	Total ears	(%)
a	10	-	14	-	0	-	24	-
a + b	47	(57)	23	(28)	12	(15)	82	(100)
a + c	36	(50)	28	(39)	8	(11)	72	(100)
a + b + c	15	(63)	5	(21)	4	(16)	24	(100)
a + d	79	(42)	47	(26)	60	(32)	186	(100)
Total	187		117		84		388	(100)

Note : a. microcephaly, b. delayed speech, c. delayed development and d. cerebral palsy

TABLE 2. DISTRIBUTION OF HEARING LOSS BASED ON AGE

Age (year)	<40dB (normal mild)	(%)	41-105dB (moderate-profound)	(%)	105dB (-) (endocochlear damage)	(%)	Total ears	(%)
<1	23	(24)	39	(41)	34	(35)	96	(100)
1-4	142	(56)	66	(26)	44	(28)	252	(100)
> 5	22	(55)	12	(30)	6	(15)	40	(100)
Total	187		117		84		388	(100)

Discussion

The risk factors for screening HL in neonates [7] are one of the following: (1) family history of hereditary childhood HL, (2) congenital perinatal infection (TORCH), (3) malformation of the head or neck, (4) birth weight < 1500 grams, (5) hyperbilirubinemia, (6) ototoxic medications, (7) bacterial meningitis, (8) 1 minute Apgar score of 0-4 or 5 minute Apgar score of 0-6, (9) mechanical ventilation longer than 5 days, and (10) stigmata of syndromes.

This study found 14 ears patients with microcephaly have moderate to profound HL. The ears of patients with microcephaly and delayed speech showed moderate to profound HL (28%) and endocochlear damage (15%). The ears of patients of microcephaly and cerebral palsy showed moderate to profound HL (26%) and endocochlear damage (32%). This result support that evaluation of hearing loss are important for patients with speech delay and cerebral palsy.[8]

The ears of patients with microcephaly and delayed development have moderate to profound HL (39%) and endocochlear damage (11%). The ears of patients with microcephaly delayed speech and delayed development have moderate to profound HL (25%) and endocochlear damage (20%). Squires *et al* (1980)[9] found that 73% Down syndrome patients had hearing impairment.

About 126 patients (65%) referred for hearing evaluation with age between 1-4 years old. Their ears have moderate to profound HL (26%) and endocochlear damage (28%). Patients with age below 1 year old had ears with endocochlear damage (35%). This finding support the recommendation to screen hearing loss in babies below 6 months of age.[1]

In conclusion, this study revealed the importance of early HL detection in microcephalic patients especially those with other handicaps such as delayed speech, delayed development and cerebral palsy.

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