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Steady-State Evoked Potential and Behavioral Hearing Thresholds in a Group of Children with Absent Click-Evoked Auditory Brain Stem Response

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Abstract
Objective: 1) To examine the distribution of behavioral hearing thresholds in a group of children who had shown no click-evoked auditory brain stem response (ABR) at maximum presentation levels. 2) To describe the relationship between the 90 Hz steady-state evoked potential (SSEP) and behavioral thresholds in these subjects.

Design: A retrospective study based on clinical findings obtained from 108 infants and young children. Each of these children had shown no recordable ABR to clicks presented at maximum levels (100 dB nHL). SSEP audiograms were obtained using AM/FM tones at the octave frequencies 250 to 4000 Hz. The results of these evoked potential assessments were compared with hearing thresholds established behaviorally.

Results: Click-ABR assessment could not differentiate between the subjects in our sample with total hearing losses and those with useful residual hearing. Although some of the ears were anacusic, more than a quarter showed residual hearing at each of the audiometric frequencies. Furthermore, at least 10% of the behavioral thresholds at each frequency fell within the moderate/severe hearing loss range. A far closer relationship was observed between SSEP and hearing thresholds. On occasions where the SSEP was absent at maximum levels, 99.5% of the ears showed either a total loss or a behavioral threshold within 10 dB of that level. When an SSEP was obtained, the hearing threshold was typically within 5 dB of the SSEP threshold.

Conclusion: The results suggested that in our group of selected subjects, the SSEP technique was able to assess ears with only minimal amounts of residual hearing. Where the brevity of the acoustic click limits both its frequency specificity and its presentation level, the modulated tones used for SSEP testing allow accurate, frequency-specific assessment at high presentation levels.

Early diagnosis of congenital hearing loss and prompt intervention has been shown to have significant long-term benefits for the development of listening, speech, and language skills in young children (Downs, 1986; Markides, 1986; Ramkalawan & Davis, 1992). This is particularly the case for infants with severe to profound hearing loss, who, without appropriate assistive devices, would have limited or no access to speech signals.

The fact that behavioral testing of hearing in very young or developmentally delayed children can be unreliable has lead, over the past few decades, to the development of assessment techniques that do not require volitional responses from the subjects (Davis, Wharrad, Sancho, & Marshall, 1991; Durieux-Smith, Picton, Edwards, & MacMurray, 1985; Moncur, 1968). Otoacoustic emission (OAE) and evoked potential assessments have emerged as the tests of choice for these populations.

OAEs are releases of sound energy within the cochlea that are recordable in the ear canal (Kemp, 1978). The presence of this emission has been shown in a number of studies to be consistent with normal or near normal hearing. OAEs are not, however, useful for estimating the audiogram in subjects with greater than mild-moderate sensory impairments because they are absent for hearing losses above 35 dB HL (transient evoked OAEs) and 50 dB HL (distortion product OAEs) (Harris, 1990; Lonsbury-Martin, Whitehead, & Martin, 1991; Nelson & Kimberley, 1992; Norton, 1993).

Of the evoked potential techniques, the auditory brain stem response (ABR) appears to be the most useful for estimation of hearing level in young children (Picton, Durieux-Smith, & Moran, 1994). The most commonly used ABR test paradigm employs acoustic click stimuli. The advantage of this stimulus lies in its brevity, which generates the synchronous neural firing in the auditory pathway required for successful measurement of the response. A number of authors have described the close relationship between hearing level and click-ABR threshold level in subjects with varying degrees of sensorineural hearing loss (Gorga, Worthington, Reiland, Beauchaine, & Goldgar, 1985; Van der Drift, Brocur, & Van Zanten, 1987).

The click-evoked ABR technique does, however, have a number of major shortcomings in the
assessment of ears with significant degrees of hearing impairment. Firstly, maximum click presentation levels are limited. Because of the acoustic click's short duration (100 μsec), the threshold advantage seen for pure tones arising from "temporal integration" is not obtained. As a result, behavioral thresholds for the acoustic click usually are found at levels between 30 dB SPL (peak) and 36 dB SPL (peak) in normally hearing subjects for presentation rates between 10 Hz and 20 Hz. (Burkard, 1984; Klein & Teas, 1978; Stapells, Picton, & Smith, 1982). Because maximum output levels for most clinical ABR systems are approximately 130 dB SPL (peak), click ABR testing usually is restricted to corrected presentation levels of less than or equal to 100 dB nHL. The possibility of residual hearing at profound levels, therefore, cannot be investigated thoroughly using the click-ABR.

The second major limitation of click-ABR testing lies in the broad frequency spectrum of the stimulus. Because the click contains acoustic energy across a wide range of frequencies, it can be difficult to determine accurately which cochlear regions are contributing to the response (Eggermont, 1982). A certain degree of frequency specificity does, however, arise from the mechanical properties of the cochlea. The click-ABR threshold has been shown in both adults and children to correlate most strongly with the best behavioral threshold in the 1000 Hz to 4000 Hz frequency range (Durieux-Smith et al., 1991; Hyde, Riko, & Malaizia, 1990; Stapells, 1989). As a result, an ear with useful low-frequency hearing but high-frequency thresholds in the severe to profound range is likely to show no ABR at maximum presentation levels and be indistinguishable from an ear that is anacusic. This effect was described by Brookhouser, Gorga, and Kelly (1990), who found that, in their sample of 196 ears that had shown no click-ABR at 100 dB nHL, behavioral thresholds could be obtained with a 500 Hz warble tone on more than 60% of occasions.

Reasonably frequency-specific stimuli such as brief tone bursts and tone pips have been employed in a number of studies to elicit the ABR (Munnerley, Greville, Purdy, & Keith, 1991; Stapells, Greville, & Martin, 1995; Stapells, Picton, Durieux-Smith, Edwards, & Moran, 1990). Although these techniques have shown some ability to investigate hearing in the low-frequency range, they, too, are restricted in their presentation levels by the necessity for stimulus brevity. Normal behavioral thresholds for tone pips, for example, are between 23 dB SPL (peak) and 29 dB SPL (peak). Maximum presentation levels for these stimuli, therefore, are limited to around 100 dB nHL (Stapells et al., 1990). Hearing losses that these limits cannot, as a result, be described accurately by tonal ABR assessment.

Steady-state evoked potential (SSEP) testing recently has become available as an objective hearing test option for young children. SSEPs are periodic scalp potentials that arise in response to regularly varying stimuli such as a sinusoidal amplitude and/or frequency modulated tones (Kuwada, Batra, & Maher, 1986; Lins, Picton, Picton, Champagne, & Durieux-Smith, 1995; Rance, Rickards, Cohen, De Vidi, & Clark, 1995; Rickards & Clark, 1984). The response is generated when the stimulus tones are presented at a rate that is sufficient to cause an overlapping of transient potentials. The energy in the resultant response is at the modulation frequency and its harmonics, allowing response detection using automatic and objective analysis protocols (Cohen, Rickards, & Clark, 1991; Jerger, Chimel, Frost, & Coker, 1986; Stapells, Makeig, & Galambos, 1987). The SSEP fulfills the two major criteria for clinical application in pediatric populations. The response is recordable at low sensation levels in sleeping and sedated subjects when modulation rates in excess of 70 Hz are used (Cohen et al., 1991; Levi, Folsom, & Dobie, 1993; Lins et al., 1996; Lins & Picton, 1995) and is reliably present in children of all ages, including neonates (Aoyagi, Kiren, Kim, Suzuki, Fuse, & Koike, 1993; Rance et al., 1995; Rickards, Tan, Cohen, Wilson, Drew, & Clark, 1994).

SSEP testing using modulated tones offers significant advantages over techniques that require short-duration stimuli. Because the tones are continuous, they do not suffer the spectral distortion problems associated with brief tone bursts or clicks. As such, they are comparatively
frequency-specific. Amplitude modulated tones, for example, contain energy only at the carrier frequency and the carrier frequency plus and minus the modulation frequency (Kuwada et al., 1986). This specificity allows testing across the audiometric range and the generation of evoked potential audiograms, which, in subjects with hearing impairments, can reflect the configuration of the loss accurately (Lins et al., 1996; Rance et al., 1995).

The continuous nature of the SSEP stimuli also offers a presentation level advantage over short-duration stimuli. The modulated tones employed to elicit the response are similar to the warble tones used in behavioral assessment. As such, the calibration corrections associated with tone bursts and clicks are not required, and the stimuli can be presented at levels as high as 120 dB HL. Investigation of ears with only minimal amounts of residual hearing is therefore possible as was demonstrated in our previous papers (Rance, Rickards, Dowell, Cohen, & Clark, 1994; Rance et al., 1995). These studies found a close relationship between SSEP and behavioral thresholds in subjects with sensorineural hearing losses ranging from mild to profound degree.

This paper examines the SSEP and behavioral threshold findings for a selected group of children who had shown no recordable ABR to acoustic clicks presented at 100 dB nHL. The first phase of the study is concerned with the levels of residual hearing found in these ears. The second phase examines the relationship between the SSEP and behavioral thresholds found in these cases.

**Method**

**Subjects**

Findings from a total of 108 infants and young children, aged 1 to 49 mo at the time of the SSEP assessment (mean age = 25 mo; median age = 24 mo) are presented in this study. The subjects were included only if ABR testing failed to show repeatable waveforms to acoustic clicks presented at 100 dB nHL.

Fifty-five of the subjects were referred to the Victorian Children's Hearing Centre (VCHC) for SSEP testing as part of their preoperative evaluation for the cochlear implant procedure. In these cases, behavioral audiometric testing carried out by audiologists in the University of Melbourne Cochlear Implant Clinic already had established the presence of profound hearing loss before the SSEP assessment. For the remaining 53 subjects, SSEP testing was performed as a part of the initial hearing loss diagnosis. These children usually were referred after a poor result on a neonatal ABR screening test. As a result, behavioral thresholds typically were obtained between 6 and 12 mo after the evoked potential assessments.

The hearing losses in each of these children appeared to be sensorineural in nature. A subject was excluded if an air-bone gap was indicated (where bone conduction results were available) or if abnormal tympanometric results were observed on any of the test occasions. For children under the age of 6 mo, tympanograms were obtained using multiple probe frequencies to reduce the possibility of artifactual results (Marchant, McMillan, Shurin, Turczyk, Feinstein, & Panek, 1986).

**Apparatus and Procedures**

**Behavioral Testing** • Behavioral audiometric thresholds were obtained by audiologists at a number of agencies including the VCHC, the Monash Medical Centre (MMC), and the Australian Hearing Services. As a result, the specific audiometric test equipment used in the testing varied by facility. All testing was, however, carried out in double-walled, sound attenuating rooms, and standard clinical test protocols were used.

Unaided and aided behavioral thresholds were established using clinical audiometers and test
techniques appropriate to the developmental level of the child (visual reinforcement audiometry/conditioned play audiometry). Threshold levels were determined using the 10 dB down, 5 dB up search technique. Unaided testing was carried out under headphones using warble tones at octave frequencies in the range 250 Hz to 4000 Hz. Maximum presentation levels were 120 dB HL at all frequencies apart from 250 Hz, which was limited to 105 dB HL.

Aided testing also employed warble tone stimuli and was carried out in the free field. The thresholds in this case were in dB SPL, and the upper limit of stimulation was at a level sufficient to elicit the maximum power output of the hearing aid (>90 dB SPL). The hearing aids were supplied and fitted by the Australian Hearing Services according to the prescription described by Byrne, Newall, and Parkinson (1990).

Where possible, complete audiograms were obtained, but the constraints of clinical testing in some cases prevented the acquisition of thresholds at each test frequency. All of the results included in this study were considered reliable by the audiologists who completed the assessments. In most cases the children were assessed over a number of test sessions, and the results were shown to be repeatable.

Evoked Potential Testing • At the time of the ABR and SSEP assessments, the children were either in natural sleep (n = 15), sedated with chloralhydrate (50 mg/kg; n = 58), or under a general anesthetic (halothane 1:1 nitrous oxide; n = 35). Responses were sought only while the subject was still and quiet.

The EEG activity for both the ABR and SSEP testing was recorded using silver-silver chloride disk electrodes placed on the forehead (+ ve) and on the earlobe or mastoid ipsilateral to the stimulated ear (- ve). A third electrode on either the contralateral mastoid or the cheek acted as a ground. Intraelectrode impedance was in all cases less than 10 kohms at 260 Hz.

ABR Measurement • ABR testing was performed at the VCHC or at the MMC. Similar procedures and test facilities were employed in each clinic.

ABR assessments at the VCHC were carried out using a system that was custom built in association with the University of Melbourne, while testing at the MMC was performed with the Biologic Navigator apparatus. The test parameters and procedures in the two clinics were identical in all respects, apart from click presentation rate, which at the VCHC was 12 Hz and at the MMC was 30 Hz. Because the effects of click rate in this range on ABR threshold have been shown to be negligible, this difference was not thought to be significant (Sininger & Don, 1989).

Responses were sought to 100 μsec acoustic click stimuli presented monaurally via Etymotic Research ER-3A tubephones at a maximum level of 100 dB nHL (Etymotic Research, 1985). The click stimuli were calibrated with a DB-0138 2-cc coupler with the sound tube connected via a dB-0138 nipple. Peak-equivalent methods were used (Glatke, 1983). A panel of 10 normal-hearing adults were tested to establish the nHL levels. The results showed that a 0 dB nHL stimulus had a peak equivalent SPL of 36 dB. As such, the maximum presentation level for the clicks was 136 dB SPL (peak).

The raw EEG signal was amplified and filtered using a band pass of 100 Hz to 3000 Hz (slope 12 dB/octave). Trials containing amplitudes in excess of 15μV were automatically rejected. A poststimulus analysis time of 12 msec was used, and 2000 samples were included in each averaged response. It is possible that the test paradigms employed in this study may not have been optimal for the recording and analysis of the ABR. The clinical systems that were used, for example, did not allow for a reduction in the high-pass EEG filter setting or an increase in the analysis time window. Such modifications may, in some cases, have allowed the recording of
responses from low-frequency cochlear regions.

Patients were included in the study only when ABRs were absent for six trials of clicks presented at 100 dB nHL. Initial testing employed a combination of condensating and rarefacing clicks, alternated to reduce the risk of stimulus artifact (two trials). In the absence of repeatable waveforms, testing then was carried out using unipolar clicks to investigate the presence of the cochlear microphonic (CM) potential (two trials for each polarity).

**SSEP Measurement** • All of the SSEP assessments were carried out at the VCHC using a custom-built evoked potential system that employed an IBM-compatible XT-type microcomputer to generate stimuli and analyze responses in the manner described by Cohen et al. (1991). The raw EEG signal was passed through a preamplifier and filtered using a band pass of 0.2 Hz to 10 kHz. The signal then underwent a Fourier analysis at the stimulus modulation frequency (90 Hz) using analog multiplication followed by low-pass filtering (Regan, 1966). Two multipliers and two low-pass filters were employed to extract both response phase and response amplitude information. The presence or absence of a response then was determined automatically from these data using a detection criterion that looked for nonrandom phase behavior. The particulars of this analysis, which is equivalent to the phase coherence technique described by Jerger et al. (1986) and by Stapells et al. (1987), are discussed in detail in Cohen et al. (1991) and Rance et al. (1995). The system calculated the probability that a set of observed phase angles could arise in the absence of a response (i.e., be the result of random EEG activity). If the probability of this occurring was sufficiently small \( p < 0.01 \) over a given period of time, a response was considered to be present and the run was terminated. A minimum of 60 samples was required before a run could be accepted, and a response was considered to be absent if the probability criterion had not been reached after 256 samples.

The test stimuli were 250 Hz, 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz tones, amplitude- and frequency-modulated at rate of 90 Hz. A modulation rate of 90 Hz was used to avoid the problems associated with eliciting the SSEP in sleeping subjects with stimuli presented at lower modulation frequencies. An amplitude modulation depth of 100% and a frequency modulation width of 10% were combined to maximize response amplitude (Cohen et al., 1991). The stimulus tones were presented via mu-metal shielded TDH-39 headphones that allowed maximum sound levels of 104 dB HL for the 250 Hz carrier and 120 dB HL for the 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz carrier frequencies. Calibration was performed using pure tones as per the AS 1591.2 standard, a B & K artificial ear (6-cc coupler), a B & K 2613 amplifier, and a B & K 2120 frequency analyzer. The introduction of modulation caused a small increase in stimulus energy that was less than 2 dB (Cohen et al., 1991).

The continuous tonal stimuli were reasonably frequency-specific. The degree of frequency modulation was limited to 10% so that the energy spectra of the stimuli were essentially contained within one critical band (Cohen et al., 1991). The combination of amplitude and frequency modulation did result in an overall shift in the energy spectrum. To compensate for this effect, the carrier frequencies were reduced by 5%.

To obtain SSEP thresholds, the level of the stimulus was decreased in 10 dB steps until the response could no longer be detected. It was then increased in 5 dB steps until the potential again was identified. Threshold was defined as the minimum level at which the response was detected. On occasions where no SSEP was obtained at the maximum presentation level, the run was repeated. Testing was carried out in this way for each of the subjects, using stimuli presented monaurally at a range of carrier frequencies. Time constraints, particularly for the subjects in natural sleep, did, however, mean that not all of the frequencies could be assessed for every child.
Results
Behavioral Thresholds in the Absence of ABR and SSEP Responses at Maximum Levels

Figure 1 shows the distribution of behavioral unaided thresholds (dB HL) obtained from the subjects at the audiometric frequencies (250 Hz to 4000 Hz). The number of ears represented in each distribution is shown in the top left of each panel. Numbers vary because at the time of data collection, complete behavioral audiograms had not yet been obtained for every child.

For 105 of the 108 subjects in this study, the lack of response on the click-evoked ABR assessment was consistent with significant sensorineural impairment. Results indicated the presence of hearing losses of at least moderate degree for each of these children. Three of the subjects showed normal or only mildly elevated behavioral thresholds. In each of these cases (which are presented separately in a later section), there was evidence of retrocochlear abnormality. The findings for these children have not been included in the group data analyses.

Many of the subjects in our sample (particularly the cochlear implant candidates) showed no residual hearing at some or all of the audiometric frequencies. There were, however, a significant number of occasions where some degree of residual hearing was found in these ears that had shown no response on click-ABR testing. This finding is described in Table 1. Although the percentage of ears with behavioral responses decreased with increasing frequency, more than a quarter of the ears (27%) still showed some residual hearing at 4000 Hz.

The absence of the click-ABR did not preclude the presence of residual hearing at severe or even moderate levels. At each of the audiometric frequencies, more than 25% of the behavioral thresholds obtained for ears with residual hearing were at levels <= 100 dB HL, and more than 10% were at levels <= 90 dB HL (see Table 2 for details). Furthermore, behavioral thresholds at
levels as low as 55 dB HL to 65 dB HL were found for each of the test frequencies.

<table>
<thead>
<tr>
<th>Frequency</th>
<th>Threshold &lt;= 100 dB HL (%)</th>
<th>Threshold &lt;= 90 dB HL (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>500 Hz</td>
<td>37</td>
<td>36</td>
</tr>
<tr>
<td>1000 Hz</td>
<td>26</td>
<td>10</td>
</tr>
<tr>
<td>2000 Hz</td>
<td>24</td>
<td>10</td>
</tr>
<tr>
<td>4000 Hz</td>
<td>20</td>
<td>10</td>
</tr>
</tbody>
</table>

The findings for ears that in addition to showing no click-ABR also showed no SSEP response at maximum presentation levels are also described in Figure 1. In this case, the behavioral result at a particular frequency was included if no SSEP could be elicited by a stimulus tone of the same frequency. There were relatively few occasions where residual hearing was found in the absence of the steady state response (see Table 1). Results varied with frequency, but for stimuli between 500 Hz and 4000 Hz, the percentage of ears with behavioral thresholds was <=21%. Behavioral responses were more commonly seen for the 250 Hz tone, but many of these are likely to be of vibrotactile rather than auditory origin (Boothroyd & Cawkwell, 1970).

Figure 1 shows the small range of behavioral thresholds that were seen where the SSEP response was absent. For all test frequencies, on each occasion where the SSEP could not be obtained (n = 365), the behavioral threshold was either absent or was found within 15 dB of the maximum presentation level. Three hundred sixty-three of the 365 results were, in fact, within 10 dB of the maximum level. The best behavioral threshold observed at frequencies between 500 Hz and 4000 Hz was 105 dB HL, and at 250 Hz was 95 dB HL.

In addition to these comparisons where the SSEP was absent at maximum levels, there were 43 occasions where the SSEP was present in the absence of any behavioral response. Because the SSEP threshold in each of these instances was seen only at levels in close proximity to the absent behavioral response (120 dB HL), it is possible that these results are a reflection of intertest variations. The SSEP and behavioral assessments were carried out in different test settings and were, in most cases, conducted many months apart.

Some of the responses also may have been the result of somatosensory rather than auditory stimulation. The majority of instances (27/43) where SSEPs were obtained in the absence of behavioral hearing thresholds occurred with low-frequency stimuli (250 Hz and 500 Hz). It is possible that at high presentation levels, low-frequency tones can produce steady-state potentials similar to those described by Snyder (1992) for vibrotactile stimulation on the hand.

There were no occasions where the SSEP threshold was seen at a level more than 10 dB better than an absent behavioral response.

**SSEP/Behavioral Threshold Comparisons**

Overall, 454 comparisons of SSEP and behavioral threshold were obtained from the 105 subjects. One hundred thirty-eight of these were at 250 Hz, 129 were at 500 Hz, 108 were at 1000 Hz, 47 were at 2000 Hz, and 32 were at 4000 Hz. There were fewer data points available for the high-frequency stimuli because many of the children had corner audiograms and showed no response for either the SSEP or behavioral assessments at maximum presentation levels. An example of the SSEP and behavioral results obtained for such children can be seen in Figure 2 (audiogram a). The results presented in audiogram b are similar to those observed for the ears in our sample that had some residual hearing. The evoked potential thresholds in this case are typical in that they slightly overestimate the hearing thresholds and closely reflect the
The relationship between behavioral and SSEP threshold for each of the ears at each of the test frequencies is shown in Figure 3. The solid lines in the distributions are the linear regression lines derived from our previous study on SSEP findings in ears with sensorineural hearing loss (Rance et al., 1995). The results for the current study are consistent with the previous findings.

The SSEP thresholds typically were obtained at close proximity to the behavioral thresholds. On 432 of the 454 occasions (99%), the SSEP threshold was seen within 20 dB of the behavioral level. In 95% of cases, the SSEP and behavioral levels were within 15 dB, and differences of less than or equal to 10 dB were found for 82% of the comparisons. There were no instance where the SSEP threshold was more than 25 dB from the observed behavioral threshold.

Difference values were established for each of the SSEP/behavioral threshold comparisons. These values were obtained by subtracting the behavioral from the SSEP level. The difference findings fit a normal distribution pattern and were similar for each frequency. Mean difference...
levels ranged from 3.1 dB to 6.3 dB for the various stimuli. The standard deviations of the mean difference levels also were similar across frequencies, varying from 6.4 dB to 8.1 dB. These findings are summarized in Table 3. The data indicate that for hearing losses in the severe to profound range, the SSEP can be recorded at levels close to behavioral threshold. This finding, in conjunction with the small spread of behavioral thresholds observed for each of the stimulus frequencies, shows that the audiogram of patients with severe-profound hearing loss can be predicted with confidence on the basis of SSEP results.

**TABLE 3. SSEP/Behavioral threshold difference values.**

<table>
<thead>
<tr>
<th>Frequency</th>
<th>Mean dB</th>
<th>SD dB</th>
</tr>
</thead>
<tbody>
<tr>
<td>250 Hz</td>
<td>1.4</td>
<td>0.7</td>
</tr>
<tr>
<td>500 Hz</td>
<td>0.3</td>
<td>0.7</td>
</tr>
<tr>
<td>1000 Hz</td>
<td>2.4</td>
<td>1.5</td>
</tr>
<tr>
<td>2000 Hz</td>
<td>3.1</td>
<td>0.4</td>
</tr>
<tr>
<td>4000 Hz</td>
<td>5.6</td>
<td>0.6</td>
</tr>
</tbody>
</table>

The Relationship between SSEP and Aided Behavioral Thresholds

Aided behavioral threshold results were available for 90 of the 108 children. Findings at 1000 Hz, 2000 Hz, and 4000 Hz are described in Table 4. The percentage of thresholds at less than or equal to 60 dB SPL was determined for the whole group and for subjects grouped according to their SSEP threshold level at each frequency. The 60 dB SPL cut-off level was selected because it represents the limit of the average long-term 70 dB SPL speech spectrum at these frequencies.

**TABLE 4. The percentage of aided behavioral thresholds at levels <=60 dB SPL for ears with no click-ABR and for ears with various SSEP levels.**

<table>
<thead>
<tr>
<th>Frequency</th>
<th>1000 Hz</th>
<th>2000 Hz</th>
<th>4000 Hz</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal ABR</td>
<td>34.0%</td>
<td>31.1%</td>
<td>30.3%</td>
</tr>
<tr>
<td>SSEP Threshold</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>120 dB HL</td>
<td>17.0%</td>
<td>30.0%</td>
<td>21.1%</td>
</tr>
<tr>
<td>110 dB HL</td>
<td>40.0%</td>
<td>30.0%</td>
<td>21.1%</td>
</tr>
<tr>
<td>100 dB HL</td>
<td>58.0%</td>
<td>40.0%</td>
<td>10.3%</td>
</tr>
</tbody>
</table>

Results varied with frequency, but for each of the test stimuli a significant number of aided behavioral thresholds were obtained at levels <=60 dB SPL. For the 161 ears tested, 54% of the thresholds at 1000 Hz, 26% of the thresholds at 2000 Hz, and 14% of the thresholds at 4000 Hz were within this range. Because each of the ears had shown no response on click-ABR testing, the results indicate that the absence of the ABR at maximum click presentation levels does not preclude the possibility of aided hearing within the speech range.

In ears that had shown no SSEP at maximum presentation levels, on the other hand, aided behavioral thresholds at levels <=60 dB SPL were uncommon. Only 12% of ears at 1000 Hz, 5% of the ears at 2000 Hz, and 2% of the ears at 4000 Hz had aided hearing within this range.

On occasions where an SSEP was obtained, the percentage of ears with aided behavioral thresholds at levels <=60 dB SPL varied with the SSEP threshold. For each of the test frequencies, fewer aided responses were obtained within this range as the level of the SSEP threshold increased. In ears where the SSEP threshold was at a level of <=105 dB HL, for example, behavioral aided thresholds were, on all occasions, obtained at <=60 dB SPL. Ears with an SSEP threshold of 120 dB HL, on the other hand, were less likely to show aided hearing within the speech range, with only 60% of the thresholds at 1000 Hz, 35% of the thresholds at
2000 Hz, and 21% of the thresholds at 4000 Hz found at ≤60 dB SPL.

Results for Children With No ABR But CM Responses

Table 5 shows findings for three subjects in the group of 108 children who had shown no ABR at maximum presentation levels. The results obtained from these children were not included in the previously described analyses. Although their lack of response to alternating clicks presented at 100 dB nHL made them indistinguishable from the other subjects in the group, the presence of the CM potential to unipolar clicks was atypical.

TABLE 5. Summary of results for three children with no identifiable ABR but cochlear microphonic responses.

The presence of the CM was confirmed in each of the subjects via control recordings carried out with the ER-3A tubephones securely clamped during stimulus presentation. This allowed verification that the potentials obtained in the unclamped condition were not due to electrical artifact.

An example of the results obtained from these children can be seen in Figure 4. The first tracings without an evident ABR waveform were obtained using alternating condensation and rarefaction clicks at 100 dB nHL. Immediately below are the traces for rarefaction clicks presented at 80 dB nHL. In this case, the CM with well-defined peaks beginning at a latency of around 1 msec can be identified clearly. The third pair of tracings shows the 180° phase shift in the CM potential that arose when clicks of opposite polarity (condensation) were used. The final tracings were obtained to rarefacting clicks presented with the tubephone clamped. In this case, the stimulus artifact still can be observed, but the attenuation of the clicks has resulted in the abolition of the CM response.

Figure 4. Averaged EEG findings for patient J.A. The top tracings show no repeatable potentials to alternating acoustic clicks presented at 100 dB nHL. The second and third tracing pairs show clear cochlear microphonic responses but absent ABR waveforms to unipolar stimuli at 80 dB nHL. The final tracings were obtained to rarefacting clicks presented with the tubephone clamped.
The behavioral thresholds obtained for these children were significantly better than those of their peers. At 4000 Hz, where the best threshold seen for the other subjects was 65 dB HL and where 95% of the thresholds were found at levels >=90 dB HL, responses for patient D.C. were obtained at 50 dB HL and for patients J.A. and C.R. were as low as 15 dB HL. Similarly, at 1000 Hz, where 95% of the behavioral thresholds were obtained at levels >=85 dB HL, thresholds for these three patients were between 15 dB HL and 45 dB HL.

The SSEP findings for these children were also unusual. Potentials were obtained in all of the cases but only at high sensation levels (see Table 5 for details). The range of observed SSEP/behavioral difference values for these cases (45 dB to 80 dB) was higher than expected given the findings of this study and previously published results (Rance et al., 1995). This inconsistency suggests that the abnormality affecting the ABR in these cases is likely to have influenced the SSEP results also.

**Discussion**

The results of this study show that, although the absence of the click-evoked ABR at maximum presentation levels is, in most cases, a good indicator of the presence hearing impairment, the ABR technique is insensitive to threshold variations within the severe to profound hearing loss range. Identical results (i.e., response absence at 100 dB nHL) were obtained in a group of young patients whose hearing levels ranged from total hearing loss to only moderate/severe impairment.

There were in our sample a significant number of ears that showed behavioral thresholds at or below the maximum click presentation level (100 dB nHL). At each of the audiometric frequencies, more than 25% of the thresholds were obtained at levels <=100 dB HL, and more than 10% of these fell within the moderate to severe hearing loss range (<=90 dB HL) (Table 2).

The click-ABR assessments were particularly insensitive to the presence of low-frequency hearing in these children. This finding may in part be due to the recording paradigm employed in the testing. Had a longer poststimulus analysis time and lower high-pass EEG filter setting been used, it is possible that responses from low-frequency cochlear regions may have been obtained in some cases. The results were, however, consistent with those of other click-ABR studies that have shown that the response threshold does not accurately reflect low-frequency hearing levels (Durieux-Smith et al., 1991; Garga et al., 1985; Stapells, 1989; van der Drift et al., 1987). At 500 Hz, 59% of the behavioral thresholds were obtained at levels less than or equal to the maximum click presentation level. An even greater proportion of the ears (97%) had behavioral responses at 250 Hz at this level. A number of these may, however, have been vibrotactile in origin. As expected, the absent click-evoked ABR was more indicative of profound hearing loss in the high-frequency range, but even at 2000 Hz and 4000 Hz a significant number of ears showed residual hearing at or below the maximum click level (2000 Hz-27%; 4000 Hz-30%).

Furthermore, many of the ears showed hearing thresholds at levels beyond the range of the click stimulus. Results varied with frequency, but behavioral thresholds were obtained to tones presented at or below 120 dB HL on between 88% (500 Hz) and 48% (4000 Hz) of occasions.

A number of studies have suggested that some of the frequency specificity limitations associated with ABR testing can be overcome by employing brieftone rather than click stimuli. Results showing reasonable behavioral/tone burst ABR threshold correlations (500 Hz to 4000 Hz) have been presented for both children and adults with normal hearing and with varying degrees of hearing loss (Munnerley et al., 1991; Stapells et al., 1995; Stapells et al., 1990). The usefulness of this technique in the assessment of subjects with severe/profound hearing loss is, however, limited by stimulus presentation level restrictions. As is the case with click stimuli, the brevity of these
short duration tones prevents testing at levels higher than approximately 105 dB nHL.

The absence of SSEP responses at maximum levels was a reliable indicator of profound or total hearing loss. The continuously modulated tones used to elicit the SSEPs could be presented at levels as high as 120 dB HL, allowing greater degrees of impairment to be evaluated. The frequency specificity of the stimulus tones also allowed assessment of residual hearing across the audiometric frequency range. On the majority (82.5%) of occasions where no SSEP response could be obtained at a particular frequency, behavioral testing indicated total hearing loss. Furthermore, for the few instances where a hearing threshold was obtained in the absence of an SSEP response, the behavioral level always was found within 15 dB of the maximum SSEP level. On 99.5% of the occasions, the threshold was, in fact, within 10 dB. This very small range of observed hearing thresholds suggests that estimates of "best possible" behavioral threshold can be made on the basis of SSEP absence with far greater confidence than is the case for an absent click-ABR result.

When an SSEP was obtained, the range of observed behavioral thresholds was also small. Overall, 82% of the SSEP thresholds were within 10 dB of the subjects' hearing levels, and 95% were within 15 dB. The close SSEP/behavioral relationship was consistent across the frequency range, with mean difference levels of approximately 5 dB obtained for each of the stimulus tones (250 Hz to 4000 Hz). This finding may be the result of recruitment, which in ears with significant sensorineural hearing loss can lead to a more pronounced increase in SSEP amplitude near threshold. This in turn allows response detection at low sensation levels. Similar results were obtained for the severe/profound ears in our previous study (Rance et al., 1995).

Recruitment also may have contributed to the small spread of observed SSEP thresholds. Standard deviation values describing the variability of SSEP/behavioral differences at each frequency were consistently small (6.4 dB to 8.1 dB). The findings show that the SSEP technique can be used to make reliable hearing level predictions for ears with hearing losses in the severe to profound range.

These findings for individual frequencies translated into accurate descriptions of the subjects' hearing losses. Results obtained from individual ears consistently showed that the configuration of the audiogram could be predicted on the basis of SSEP thresholds. The relationship between the evoked potential and behavioral audiograms shown in Figure 2 is typical of that observed for our subjects, with the SSEP thresholds only slightly overestimating the behavioral levels and mirroring the audiogram configuration. Results such as these can provide a basis for early intervention. Hearing aid fittings with frequency response characteristics based on SSEP findings were, in fact, successfully carried out on a number of our subjects for whom behavioral results were not yet available.

Another treatment option for children with severe/profound hearing loss that has become available in recent years is the cochlear implant. Candidacy for cochlear implantation has, over the past decade, become a complicated issue. Where initially only subjects with total hearing losses were implanted, technical advances and promising clinical results have meant that children with small amounts of residual hearing now may be considered (Dowell, Blamey, & Clark, 1995; Osberger, 1995). The age at which children can be implanted also has reduced dramatically in recent times to the point where 1 and 2 yr olds now are considered routinely. For these children, the amount of residual hearing becomes the primary criterion for candidacy because speech perception testing (the major criterion for adults and older children) usually is not possible. The dividing line between suitable and unsuitable children is currently a matter of some debate and varies significantly between implant centers. Whatever specific criterion is employed, the decision must ultimately be based on the amount of access to speech that the child is afforded by optimally fitted hearing aids. In young hearing-impaired children, this is best described by aided
behavioral threshold results, and for children who do not always respond reliably in the
behavioral test situation, SSEP assessment can provide an estimate of likely aided behavioral
thresholds.

As was discussed previously, the absence of the SSEP at maximum presentation levels was a
strong indicator of total, or near total, hearing loss for the subjects in our group. This finding
also was reflected in the aided behavioral threshold results, where SSEP absence was, in most
cases, consistent with the absence of useful aided hearing. Only 12% of the ears at 1000 Hz, 5%
of the ears at 2000 Hz, and 2% of the ears at 4000 Hz showed aided behavioral thresholds at
<=60 dB SPL. As such, SSEP absence at these frequencies, although not precluding the
possibility of aided hearing within the normal speech range, makes such a result very unlikely.
The same could not be said for the ears that had only the click-ABR absent. In this case, more
than half of the ears (54%) had 1000 Hz aided behavioral thresholds at levels <=60 dB SPL.
Even in the higher audiometric frequencies, a significant proportion of the ears showed aided
thresholds within the speech range (2000 Hz-26%; 4000 Hz-14%). Clearly, just as the
click-ABR technique cannot accurately quantify hearing losses in the severe to profound range,
it cannot be used to differentiate between ears that will and will not have useful aided hearing.

The presence of the SSEP in our subjects was consistent with an increased likelihood of useful
aided hearing. For each of the test frequencies, the probability of obtaining an aided behavioral
threshold within the speech range increased as SSEP threshold decreased. All of the ears with
SSEP thresholds at <=100 dB HL gave aided behavioral responses within this region, and even
for ears with an SSEP threshold at 120 dB HL, there was a greater than 20% chance of obtaining
an aided threshold at <=60 dB SPL (Table 4). As such, objective results obtained using the SSEP
technique can provide an important safeguard in the cochlear implant selection process, alerting
clinicians to the possibility (or probability for SSEP thresholds <=115 dB HL) that an ear may
have useful aided hearing.

There were in our sample three children for whom no click-evoked ABR could be obtained
despite good levels of residual hearing. The presence of the CM proved a useful way of
identifying these cases. CM responses were elicited in each of these subjects during the ABR
assessment using unipolar clicks. An example of this result can be seen in Figure 4. The presence
of the CM response (which is preneural) in the absence of ABR waveforms points to a
dysfunction in the synaptic or postsynaptic areas of the auditory system. As such, these children
appear to be similar to those described by Starr, Picton, Sininger, Hood, and Berlin (1996).
Disruption of neural synchrony has been postulated as a likely cause for the pattern of results seen in such
cases (Kraus, Ozdamar, Stein, & Reed, 1984, Sininger, Hood, Star, Berlin, & Picton, 1995; Starr et al., 1996). ABR
extraction from the EEG signal, achieved by averaging, requires precise synchrony of neural
firing for response definition. Even minor variations (<0.5 msec) in the timing of neural
discharges after each stimulus can make the ABR unrecognizable.

The SSEP, on the other hand, may not require the degree of neural synchrony needed for
identification of transient waveforms. Successful recording of the potential does require that the
auditory system produce a response that is phase locked to the modulation envelope of the
stimulus. The unusually poor SSEP results obtained for these three children would suggest that
even this level of function was compromised.

The incidence of auditory disorders of this type is unclear at this stage. The number does,
however, appear to be significant. Three of the 53 cases (6%) in our diagnostic group were
identified Kraus et al. (1984) found an even higher ratio, with approximately 15% of their patients
who had shown absent or abnormal click-evoked ABRs having behavioral thresholds in the
normal to moderate hearing loss range.
Identification of these children is essential if appropriate management strategies are to be implemented. A number of authors have suggested that the fitting of hearing aids has not proven beneficial and is potentially damaging in such cases (Kraus et al., 1984; Sinner et al., 1995). Two of the three subjects in our sample were fitted binaurally with low gain aids. Both appeared to obtain little benefit, had tolerance problems, and subsequently rejected their hearing aids. The third child who had shown normal behavioral thresholds was not aided.

The findings for these children should reinforce the need for caution when making hearing loss diagnoses solely on the basis of evoked potential findings. Results obtained for both the alternating click ABR and SSEP assessments for these children (who subsequently were shown to have behavioral thresholds in the normal to moderate hearing loss range) were indistinguishable from those seen for children with severe/profound sensorineural hearing losses. Clearly, the CM findings in these instances helped identify the children whose cochlear function was better than suggested by the ABR and SSEP results. It is for this reason that testing with unipolar clicks is recommended for any subject for whom absent or abnormal waveforms are obtained in clinical ABR testing.

In summary, the findings of this study showed that in our group of selected subjects, the absence of the click-ABR did not preclude the presence of residual hearing at useful levels. Furthermore, the results indicated that the SSEP technique was able to accurately describe and quantify hearing losses in ears with only minimal amounts of residual hearing.

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