Newborn Hearing Screens May Give a False Sense of Security

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Karrie Cuttler*
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Abstract

Certain risk factors result in a higher incidence of delayed-onset hearing loss. Ten subjects who passed auditory brainstem response birth screenings and later returned with bilateral sensorineural hearing loss are examined in this report. Although not all subjects in this report had respiratory distress, this study reveals a high correlation between mechanical ventilation and delayed-onset hearing loss.

Key Words: Late-onset progressive hearing loss, sensorineural hearing loss

Abbreviations: ABR = auditory brainstem response, ECMO = extracorporeal membrane oxygenation, JCIH = Joint Committee on Infant Hearing, MAS = meconium aspiration syndrome, NICU = neonatal intensive care unit, OAE = otoacoustic emissions, PFC = persistent fetal circulation, PPHN = persistent hypertension of the newborn, RDS = respiratory distress syndrome

The prevalence of delayed-onset progressive hearing loss in children who previously passed a newborn birth screening is uncertain at this time because universal infant birth screening is relatively new. The purpose of this article is to alert examiners and related professionals to the risk of delayed-onset progressive hearing loss. A passed hearing screening at birth may give the false impression that no further follow-up is required, therefore delaying the age of diagnosis indefinitely. The time of onset of hearing loss is often unclear and requires careful monitoring.

In this study, risk factors that appear relevant to late-onset progressive hearing loss are persistent pulmonary hypertension of the newborn (PPHN), family history of hearing loss, prematurity, and mechanical ventilation. A strong association was observed between mechanical ventilation and late-onset progressive pediatric hearing loss. Although these factors have long been known as risk factors for hearing loss, it has not been evident that the hearing loss may not be apparent immediately and therefore will not be revealed on the initial birth screening.

SUBJECTS AND METHOD

Ten children were identified in our clinic with delayed-onset hearing loss after passing the newborn automated auditory brainstem response (ABR) screening. Nine of the subjects were neonatal intensive care unit (NICU) survivors, whereas one was a well-baby nursery patient. The NICU patients were screened by our staff, whereas the newborn nursery patient was screened by an audiologist from another facility at a different local hospital. Nine subjects were male and one was female. Although several of the subjects had severe respiratory distress syndrome (RDS), none received extracorporeal membrane oxygenation (ECMO) treatment. ECMO subjects are reported separately and with similar findings (Mann and Adams, 1998).

Retrospectively, medical records were examined for all of the subjects to seek precipitating factors related to hearing loss (Table 1). Persistent fetal circulation (PFC), bacterial meningitis subsequent to surgery for gastroschisis, prematurity with prolonged and mechanical ventilation, and mechanical ventilation with ototoxic exposure were found in eight of the subjects, all of whom had extended NICU stays. For

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Table 1 Relevant Subject Data

<table>
<thead>
<tr>
<th>Subject</th>
<th>Newborn or NICU</th>
<th>Initial Diagnosis</th>
<th>Gestational Age at Birth (wk)</th>
<th>Gender</th>
<th>Length of Mechanical Ventilation (d)</th>
<th>Age Hearing Loss Confirmed</th>
<th>Rehospitalized Later</th>
</tr>
</thead>
<tbody>
<tr>
<td>JW</td>
<td>NICU</td>
<td>Extreme prematurity</td>
<td>26</td>
<td>Male</td>
<td>77</td>
<td>5 yr, 6 mo</td>
<td>Age 3 yr for head injury</td>
</tr>
<tr>
<td>KA</td>
<td>NICU</td>
<td>PPHN</td>
<td>40</td>
<td>Male</td>
<td>6</td>
<td>4 yr</td>
<td>No</td>
</tr>
<tr>
<td>FC</td>
<td>NICU</td>
<td>Gastrosehisis</td>
<td>37</td>
<td>Male</td>
<td>4</td>
<td>2 yr, 8 mo</td>
<td>No</td>
</tr>
<tr>
<td>CC</td>
<td>NICU</td>
<td>MAS</td>
<td>42</td>
<td>Male</td>
<td>0</td>
<td>2 yr, 8 mo</td>
<td>No</td>
</tr>
<tr>
<td>SG</td>
<td>Newborn Nursery</td>
<td>Healthy</td>
<td>40</td>
<td>Female</td>
<td>0</td>
<td>2 yr, 4 mo</td>
<td>No</td>
</tr>
<tr>
<td>RM</td>
<td>NICU</td>
<td>PPHN</td>
<td>36</td>
<td>Male</td>
<td>14</td>
<td>1 yr, 6 mo</td>
<td>No</td>
</tr>
<tr>
<td>ZW</td>
<td>NICU</td>
<td>PPHN</td>
<td>26</td>
<td>Male</td>
<td>22</td>
<td>1 yr, 6 mo</td>
<td>Repeatedly for respiratory distress</td>
</tr>
<tr>
<td>JW</td>
<td>NICU</td>
<td>MAS</td>
<td>39</td>
<td>Male</td>
<td>10</td>
<td>1 yr, 4 mo</td>
<td>No</td>
</tr>
<tr>
<td>TW</td>
<td>NICU</td>
<td>PPHN</td>
<td>38</td>
<td>Male</td>
<td>28</td>
<td>11 mo</td>
<td>No</td>
</tr>
<tr>
<td>JS</td>
<td>NICU</td>
<td>PPHN</td>
<td>37</td>
<td>Male</td>
<td>14</td>
<td>9 mo</td>
<td>No</td>
</tr>
</tbody>
</table>

Table 2 Previous Reports of Sensorineural Hearing Loss in PPHN or RDS Subjects and Age at Hearing Test/Confirmation of Loss

<table>
<thead>
<tr>
<th>Authors</th>
<th>Subjects with PPHN</th>
<th>Prevalence of Hearing Loss (%)</th>
<th>Age at Hearing Test Confirmation of Loss</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sell et al (1985)</td>
<td>40</td>
<td>20 (N = 8)</td>
<td>2.3–4.3 yr</td>
</tr>
<tr>
<td>Naulty et al (1986)</td>
<td>11</td>
<td>27 (N = 3)</td>
<td>3–6 mo</td>
</tr>
<tr>
<td>Nield et al (1986)</td>
<td>7</td>
<td>100 (N = 7)</td>
<td>13–48 mo</td>
</tr>
<tr>
<td>Hendricks-Munoz and Walton (1988)</td>
<td>40</td>
<td>52.5 (N = 21)</td>
<td>4 mo–2 yr</td>
</tr>
<tr>
<td>Walton and Hendricks-Munoz (1991)</td>
<td>51</td>
<td>37 (N = 19)</td>
<td>2 mo–3 yr</td>
</tr>
<tr>
<td>Marron et al (1992)</td>
<td>27</td>
<td>0 (N = 7)</td>
<td>10 mo–6 yr</td>
</tr>
<tr>
<td>Kawashiro et al (1994)</td>
<td>25</td>
<td>32 (N = 8)</td>
<td>5 mo–3 yr</td>
</tr>
<tr>
<td>Kaga et al (1997)</td>
<td>23</td>
<td>34.7 (N = 8)</td>
<td>6 mo–3 yr</td>
</tr>
<tr>
<td>Konkle and Knightly (1993)</td>
<td>2</td>
<td>100 (N = 2)</td>
<td>2 yr, 6 mo</td>
</tr>
</tbody>
</table>

One subject, the only risk factor presented was mild meconium aspiration syndrome (MAS), requiring only 48 hours in NICU and no mechanical ventilation before discharge. The one female subject did not require ventilation, was a patient in the newborn nursery for only 24 hours, and reportedly had a distant cousin with hearing loss.

Many investigators have studied the prevalence of hearing loss in patients with PPHN (also known as PFC), but hearing data at the time of discharge from the hospital are not given for most of these late identified subjects. See Table 2 for previously reported prevalence of hearing loss in PPHN and RDS survivors and age of detection of hearing loss.

RESULTS

Of the 10 subjects with bilateral sensorineural hearing loss, 3 demonstrated a profound loss, 2 moderate sloping to profound, 2 moderate sloping to severe, 1 moderate flat, and 2 mild flat configuration. All 10 subjects presented with hearing loss that impaired speech and language development and would be considered educationally handicapping. Hearing losses were confirmed using behavioral audiology and ABR techniques. Degree and prevalence of bilateral sensorineural hearing losses for this study are noted in Table 3.

DISCUSSION

Despite early detection efforts of universal hearing screening programs, an insidious hearing loss may go undetected if late onset is not monitored aggressively. Newborn screening is not enough; serial follow-up testing is needed to detect hearing loss at the earliest stage of development (Mann and Adams, 1998). It is
important to recognize that a passed hearing screening at birth may not be sufficient information to preclude the possibility of pediatric hearing loss, particularly in certain high-risk cases. PPHN, meningitis, family history, prematurity, mechanical ventilation, and ototoxic exposure should be considered high-risk factors, which require intensive audiologic follow-up. Significant prevalence of delayed-onset progressive hearing loss is a concern presented by numerous researchers when investigating hearing loss (Naulty et al, 1986; Nield et al, 1986; Hendricks-Munoz and Walton, 1988; Walton and Hendricks-Munoz, 1991; Cheung and Robertson, 1994; Lund et al, 1994; Kawashiro et al, 1996; Fujikawa et al, 1997; Mann and Adams, 1998).

The large number of cases in this series with a history of PPHN is consistent with published evidence that PPHN is a significant risk factor for delayed-onset hearing loss. Although age of onset has not been clear in all cases, there is a high prevalence of hearing loss associated with this disorder in all studies except Marron et al (1992) (see Table 2). PPHN often results in the need for mechanical ventilation due to severe respiratory distress. Oxygen deprivation may be responsible for increased risk of late-onset hearing loss. Progressive degeneration of hair cells following hypoxia in chinchillas was reported by Shirane and Harrison in 1987. Likewise, however, newborns experiencing RDS and PPHN have been reported to develop hearing loss after hospital discharge (Konkle and Knightly, 1993; Kawashiro et al, 1994, 1996; Kaga et al, 1995, 1997; Mann and Adams, 1998). It is interesting that 9 of the 10 subjects were male, which leads to the question “Is male gender a stronger indicator of late-onset hearing loss?”

It is also important to be concerned if high-risk children present with otitis media. In this study, 4 of the 10 subjects suffered from chronic otitis media, which contributed to the delayed diagnosis. Parents presented concerns about the hearing loss to their pediatricians; subsequently, the physician recommended deferring audiologic evaluation until the middle ear condition resolved, further delaying the rehabilitation process. Several neonatal indicators may lead to a progressive hearing loss; thus, there is a definite need for follow-up measurements, regardless of the results of the initial birth screening.

Interim factors that may have contributed to the development of sensorineural hearing loss are difficult to document in a retrospective study (Nield et al, 1986). Just as in Nield’s study, factors present in the case histories presented appear to substantiate the results of the discharge ABR and indicate that hearing was present initially, ruling out ABR false negatives. Several families reported that the children began using words and later stopped. Parents seemed to be aware of when the speech development regressed but typically did not bring their children for audiologic evaluation until recommended by their physician, often much later than the point at which initial concern developed. One mother reported that her son stopped using words soon after the age of 1 year but did not have audiologic assessment until the age of 4 years.

Clearly, there is a significant association between mechanical ventilation and delayed-onset hearing loss. In this series of patients, 80 percent of the subjects were ventilated. New respiratory therapies may save the lives of newborns suffering with severe oxygen deprivation; however, a side effect may be a progressive hearing loss in some of the survivors (Lasky et al, 1998).

Only 2 of the 10 subjects were rehospitalized or seriously ill after discharge from NICU or newborn nursery. The two rehospitalizations were for head injury and respiratory distress. Although the nature of the rehospitalizations may be consistent with the development of hearing loss for these 2 patients, the initial medical treatment at birth is a more probable cause. Detection and habilitation efforts were significantly delayed in all 10 subjects, resulting in severe speech and language disorders.

A major concern is that the 10 children in this report passed the initial ABR birth screening; thus, follow-up recommendations were not heeded. Parents and physicians were informed in writing of the risk of late-onset hearing loss, even with passing screening results; however, the initial passing results may have given a false sense of security, which delayed the diagnosis of hearing loss. Under the 1994 Joint Committee on Infant Hearing (JCIH) recommendations, most of the
subjects in this study should have been followed for delayed-onset hearing losses. The fact that they were not denotes problems in the follow-up system that must be corrected. The importance of continued audiologic monitoring after discharge is evident from the cases presented here.

Audiologic follow-up is imperative, and late-onset progressive factors such as family history of hearing loss, RDS, PPHN, MAS, and mechanical ventilation cannot be overlooked. Another late-onset progressive factor not observed in this study is cytomegalovirus. Although prolonged mechanical ventilation was identified as a risk factor in the 1990 and 1994 JCIH statements, it was not included in the 1994 indicators for delayed-onset hearing losses. The JCIH 2000 statement expands risks factors for delayed-onset hearing loss, including PPHN and ECMO, but does not include mechanical ventilation per se. JCIH also did not give a recommendation about the frequency of monitoring.

In summary, when working to reduce the average age of detection of hearing loss, we must be sure to monitor for delayed onset. We recommend monitoring every 3 to 6 months following discharge. Sell et al (1985) and Nield et al (1986) reported initial identification and confirmation of sensorineural hearing loss as late as 4 years of age. One of our subjects was not identified until the age of 5 years and 6 months, when the school referred for audiologic assessment.

With age of detection sometimes as late as 4 or 5 years, the question arises as to when routine follow-up ends for extremely high-risk children who pass the initial screening or when follow-up should end for children without risk factors for delayed-onset hearing loss.

Audiologists alone cannot sufficiently monitor all at-risk children; thus, it is important to persevere in our efforts to educate physicians, nurse practitioners, nurses, and families about early detection of hearing loss and the risk of progressive or delayed-onset hearing loss. The efficacy of newborn screening depends, in part, on identifying newborns at risk for developing a hearing loss after hospital discharge. Identifying risk factors for progressive losses should be a high priority for those concerned with newborn screening (Lasky et al, 1998).

One future direction for research in this area is the study of otoacoustic emission (OAE) screenings and their role in early detection of late-onset progressive hearing loss. The children in this study were screened by ABR and passed at discharge time; however, the examiners question if an OAE screening would have given early insight into the cases. Recently, screening programs have incorporated OAE screenings alone or as part of a two-step screening process. Abnormal OAE screenings are often followed by ABR screenings. If OAE results are abnormal and ABR findings are normal, the screening results are considered to be a pass. The series of patients presented here bring several questions to mind. For example, would the OAE screening have been normal or abnormal for the children studied? Would an abnormal OAE screening have alerted the examiner to outer hair cell damage? Using current National Institutes of Health guidelines, an infant who fails an OAE and passes an ABR is considered to have normal peripheral auditory functioning, despite the fact that abnormal OAE findings can be consistent with early onset of hearing loss in cases of ototoxicity and outer hair cell damage. If OAE testing can detect cochlear dysfunction before it is evident on behavioral/pure-tone audiogram, can OAE also detect a cochlear hearing impairment before it is evident on ABR? Is OAE sensitive enough to be an early predictor of late-onset progressive pediatric hearing loss?

Another avenue for research consideration is the use of OAE in cases similar to the ones presented to detect children with auditory neuropathy. Although none of the subjects presented here are known to have neuropathy, many of the risk factors discussed are consistent with the potential diagnosis of auditory neuropathy. OAE screenings are helpful in differentiating cochlear (or sensory) hearing loss from true neural hearing loss. There is a current debate on whether the ABR is a more effective tool for detecting auditory neuropathy and should be used exclusively to screen the high-risk NICU population. Does this method disregard the advantages of OAE screening? Which is more prevalent, the risk of late-onset progressive hearing loss or the risk of auditory neuropathy? Is one screening method best, or should high-risk infants receive both types of screenings? These and many other questions will be answered as universal infant screening efforts continue.

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REFERENCES

Newborn Hearing Screens/Mann et al


