Delayed-Onset Hearing Loss in Respiratory Distress Syndrome: Case Reports

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Abstract

The pre-, peri-, and postnatal histories for two infants who suffered respiratory distress syndrome (RDS) are presented. Each infant was diagnosed with RDS within 24 hours after birth, placed on high-frequency jet ventilation, and passed auditory brainstem response (ABR) screening prior to hospital discharge. Both infants were enrolled in a neonatal follow-up program with no report of hearing loss during the first year of life. At 2.5 years of age, each infant was found to have severe-to-profound sensorineural hearing loss. Neither infant suffered kidney failure, rubella, cytomegalovirus, or genetic involvement. Audiologic monitoring after hospital discharge was not performed because each infant passed the ABR screen. Thus, the date of onset and/or progression of hearing loss are unknown. Until additional research findings are available, we recommend that the hearing of any infant diagnosed with RDS be monitored frequently until 3 years of age.

Key Words: Auditory brainstem response (ABR), hearing loss, infant, respiratory distress syndrome (RDS)

Respiratory distress syndrome (RDS), or hyaline membrane disease, is the most common respiratory difficulty encountered by newborns. RDS accounts for 30 percent of all neonatal deaths in the United States and 50 to 70 percent of deaths among premature infants (Rossetti, 1986). Although RDS is most common in premature infants, susceptibility depends more on lung maturation than gestational age. To our knowledge, the incidence of hearing loss associated specifically with RDS is unknown.

The following is a summary of two infants who demonstrate remarkably similar pre-, peri-, and postnatal histories. Both infants suffered RDS within 24 hours after birth, required high-frequency jet ventilation, passed auditory brainstem response (ABR) screening prior to hospital discharge, and developed normally with no report or indication of hearing loss until at least 1 year of age. Both infants, however, were subsequently found to have severe-to-profound sensorineural hearing losses at 2.5 years of age.

CASE REPORTS

Case 1

TF is a 4-year-old girl who currently suffers a bilateral sensorineural hearing loss. She was the product of a 36-week pregnancy, delivered via stat cesarean section for preeclampsia, and weighed 3000 g at birth. Apgar scores were 7 at both 1 and 5 minutes. Maternal medical history was remarkable for asthma, gestational diabetes, and toxemia.

Two hours after birth, the baby was intubated and placed on conventional ventilatory support for RDS. TF did not respond to conventional ventilation, and on day 2 of life was transferred to the infant intensive care unit (IICU) at the Children's Hospital of Philadelphia (CHOP). Upon admission to the CHOP, she was immediately placed on high-frequency jet ventilation, which was continued until day 12 of life, when she was weaned to oxygen. During this period, she developed hyperbilirubinemia, which was controlled with a 7-day course of phototherapy. TF also received a 7-day course of ampicillin and gentamicin for possible sepsis, although a positive culture was never obtained. Persistent fetal circulation syndrome was ruled out.

On day 25 of life, 2 days prior to discharge from the hospital, TF passed an ABR screening...
conducted at 40 dB nHL. The remaining 2 days of her hospitalization were uneventful, and she was discharged home on oxygen and theophylline. She was weaned completely from oxygen approximately 2 months later.

After hospital discharge, TF was enrolled in the CHOP Neonatal Follow-up Program. Regular medical check-ups and parental interviews conducted at this clinic by the neonatal follow-up team failed to identify any concerns regarding overall development, specifically as related to hearing sensitivity or speech/language development. She was released from the program at 1 year of (corrected) age.

When she was 2.5 years old, after being enrolled in a day care program for approximately 1 year, her teachers raised questions regarding her inconsistent responses to verbal commands. At the same time, her parents were becoming concerned about her speech articulation skills. Hence, her parents contacted the Neonatal Follow-up Program and a referral was made to the Division of Medical Audiology of the Children's Seashore House.

The findings of an audiologic assessment revealed hearing sensitivity for each ear to be within normal limits for octaves 250 and 500 Hz; however, there was a bilaterally precipitous hearing loss for octaves 1000 Hz and above. Pure-tone air-conduction thresholds for the right ear were obtained at 50, 80, and 95 dB HL for 1000, 2000, and 4000 Hz respectively. Thresholds for the left ear did not differ by more than 5 dB at any test frequency, as compared to those for the right ear. A subsequent speech/language evaluation revealed no significant language delay, although articulation was felt to be poor. Currently, TF uses binaural amplification and is mainstreamed to kindergarten. Her hearing loss has remained stable for the past 2 years.

**Case 2**

JF is a 6-year-old girl with a bilateral sensorineural hearing loss. She was a 34-week gestational age infant delivered via stat cesarean section for preeclampsia. Birth weight was 3600 g. Apgar scores were 8 and 8. Maternal history was remarkable for class C diabetes (e.g., duration and/or onset between 10 and 19 years). JF was diagnosed with RDS and placed on conventional ventilation.

She was maintained on conventional ventilation for the next 12 days; however, during this time, she developed several pneumothoracies that required chest tubes. As a result, she was transferred from her community hospital to the ICU at the CHOP at 12 days of life. Upon admission to the CHOP, JF was placed on high-frequency jet ventilation, which was continued until 19 days of life, when respiratory support was returned to a conventional ventilator. She was weaned to oxygen over the next 11 days. During her inpatient stay at the CHOP, she received a 7-day course of ampicillin and gentamicin. Vancomycin was added to this regime and was continued until all chest tubes were removed. She suffered transient hyperbilirubinemia, which was controlled without treatment. There was no diagnosis for persistent fetal circulation syndrome. On day 64 of life, 8 days prior to hospital discharge, she passed an ABR screening conducted at 40 dB nHL. The remainder of her admission was uneventful, and she was discharged home on oxygen, Lasix, and theophylline.

JF was followed routinely after discharge as an outpatient in the CHOP Neonatal Follow-up Program until she was 1 year old (corrected age). During this period, general development was normal, and no concerns were recorded regarding hearing, speech, or language skills.

Approximately 7 months after discharge from the program, JF's father contacted the coordinator of the program and expressed concern regarding JF's speech development. The father was counseled regarding speech and language stimulation and was invited to call again if his concerns persisted. No further anxiety was expressed until the family again contacted the program 5 months later and disclosed that they were worried about JF's hearing. At that time, JF was referred to the Division of Medical Audiology for an audiologic assessment.

Audiologic findings revealed a bilateral sensorineural hearing loss, with hearing sensitivity for each ear between 70 and 80 dB HL at octaves 250 and 500 Hz, and 90 dB HL or poorer at 1000, 2000, and 4000 Hz. JF was fitted with binaural amplification and enrolled in a school for the hearing impaired. Her hearing loss has remained unchanged for the past 2.5 years, and her academic and psychosocial progress currently is reported as satisfactory.

**COMMENT**

As stated at the outset of this report, there are a number of similarities in the case histories of these two patients. Table 1 summarizes the pre-, peri-, and postnatal factors common to each patient. As can be seen, both infants...
were delivered by stat cesarean section approximately 1 month preterm, because of preeclampsia. Apgars at 1 and 5 minutes were similar for both infants; each developed RDS within the first 24 hours after birth, each failed conventional ventilation, and the respiratory difficulties of both were successfully managed by high-frequency jet ventilation. Each infant passed an ABR screen and neither displayed delayed development during the first year of life.

In addition, neither patient suffered cytomegalovirus, rubella, kidney failure, or genetic involvement commonly associated with progressive sensorineural hearing loss. Although not specifically stated in their medical charts, it is probable that the infants suffered bronchopulmonary dysplasia, since they were both discharged from the hospital on oxygen. We are not aware, however, of any reports linking bronchopulmonary dysplasia to progressive or delayed-onset hearing loss. Both infants received drugs known to be potentially ototoxic, but these were administered in clinical dosages considered safe with respect to hearing loss. Moreover, both infants passed an ABR screen conducted at least 2 weeks following termination of the drug regime.

Based on these observations and the medical/management histories from the Neonatal Follow-up Program, it is our impression that both of the infants described in this report had essentially normal hearing at the time they were discharged from the hospital and that their hearing remained normal over the first year of life. We suspect that the hearing impairments documented by auditory assessment conducted at approximately 2.5 years of age developed either as progressive losses beginning sometime after the first year of life or as sudden impairments of delayed onset. Actual age at onset or rate of progression, however, remains unknown, since audiologic monitoring was not performed on either infant because each passed an ABR screen.

The cause of the hearing losses described in this paper cannot be specified. It is important to stress, however, that progressive and/or delayed-onset hearing loss has been reported for infants who suffered other forms of respiratory distress (Nield et al, 1986), especially persistent pulmonary hypertension of the newborn (PPHN) (Walton and Hendricks-Munoz, 1991). Clearly, PPHN and RDS are two very different respiratory disorders, yet they share common sequelae,

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**Table 1** Comparison of Common Findings in the Pre-, Peri-, and Postnatal Histories for the Two Infants Summarized in this Report

<table>
<thead>
<tr>
<th>Period</th>
<th>TF</th>
<th>JF</th>
</tr>
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<tbody>
<tr>
<td>Prenatal</td>
<td>Diabetes (gestational)</td>
<td>Diabetes (Class C)</td>
</tr>
<tr>
<td></td>
<td>Preeclampsia</td>
<td>Preeclampsia</td>
</tr>
<tr>
<td></td>
<td>Born @ 36 wk gestational age (stat c-section)</td>
<td>Born @ 34 wk gestational age (stat c-section)</td>
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<tr>
<td></td>
<td>Apgars – 7/7</td>
<td>Apgars – 8/8</td>
</tr>
<tr>
<td></td>
<td>RDS within 24 hr</td>
<td>RDS within 24 hr</td>
</tr>
<tr>
<td></td>
<td>Failed conventional ventilation (2 days)</td>
<td>Failed conventional ventilation (12 days)</td>
</tr>
<tr>
<td></td>
<td>Jet Ventilation (10 days)</td>
<td>Jet Ventilation (7 days)</td>
</tr>
<tr>
<td></td>
<td>Clinical dose gentamicin, ampicillin</td>
<td>Clinical dose gentamicin, ampicillin, vancomycin</td>
</tr>
<tr>
<td></td>
<td>Passed ABR screen (day 25)</td>
<td>Passed ABR screen (day 64)</td>
</tr>
<tr>
<td></td>
<td>Discharged (day 27)</td>
<td>Discharged (day 72)</td>
</tr>
<tr>
<td>Postnatal</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>NF-U* neg. at 1 year (30 mo)</td>
<td>NF-U* neg. at 1 year (21 mo)</td>
</tr>
<tr>
<td></td>
<td>Initial concern expressed (30 mo)</td>
<td>Initial concern expressed (21 mo)</td>
</tr>
<tr>
<td></td>
<td>Hearing loss identified (30 mo)</td>
<td>Hearing loss identified (26 mo)</td>
</tr>
</tbody>
</table>

*NF-U = CHOP Neonatal Follow-up Program.
such as neonatal hypoxia, which has been associated with cochlear insult (Sohmer et al, 1989). It is difficult, however, to generalize such observations to account for either progressive or delayed-onset hearing loss.

Finally, it should be noted that neither patient would have been identified as “at risk” for hearing loss according to the high-risk register advocated by the Joint Committee on Infant Hearing (1982), which was commonly in use at the time these patients were hospitalized. Conversely, both patients would be considered “at risk” according to the current Joint Committee on Infant Hearing (1991) high-risk register, since each required more than 10 days of mechanical ventilation. Both infants were screened for auditory function as they were about to graduate from our ICU, and both children passed their respective screenings. It may be argued, therefore, that screening programs designed to identify hearing loss in the neonatal nursery need to be acutely aware that a proportion of those infants who pass the screen will develop substantial hearing loss within the first 3 years of life. Regardless of the programmatic approach to the initial screening process (i.e., mass screening of all newborns, as compared to screening only ICU graduates or only those “at risk” for hearing loss), continued audiologic assessments at a minimum of 6-month intervals appear warranted for certain groups of high-risk infants.

Infants with PPHN have already been identified as a subgroup requiring audiologic follow-up. Based on the summary provided in this report, it appears that this subgroup should be expanded to include neonates diagnosed with RDS, regardless of their birthweight or maturational level. Until research findings delineate further the relationship between neonatal respiratory disorders and hearing loss, the hearing status of any infant diagnosed with RDS should be monitored frequently until 3 years of age.

REFERENCES


