Abstract
This study investigated the characteristics of hearing loss in children with ventriculoperitoneal (VP) shunted hydrocephalus. Twelve hydrocephalic children with patent VP shunts participated. The etiology of the hydrocephalus was either intraventricular hemorrhage or spina bifida. Audiometric examination included pure-tone air conduction thresholds, tympanometry, contralateral and ipsilateral acoustic reflex thresholds (ARTs), and distortion product otoacoustic emissions (DPOAEs). A unilateral, high-frequency, cochlear hearing loss was found in the ear ipsilateral to the shunt placement in 10 (83%) of the 12 shunt-treated hydrocephalic children. No hearing loss was observed in the ear contralateral to shunt placement. Based on the pure-tone audiometric findings, coupled with the decrease in DPOAE amplitude in the shunt ear, the hearing loss appears to be cochlear in nature. We suggest that cochlear hydrodynamics are disrupted as the result of reduced perilymph pressure, a consequence of cerebrospinal fluid (CSF) reduction due to the combined effects of a patent shunt and a patent cochlear aqueduct. In addition, a concomitant brain stem involvement is evidenced in the ART pattern, possibly produced by the patent shunt draining the CSF from the subdural space, resulting in cranial base hypoplasia.

Key Words: Cochlear aqueduct, hearing loss, hydrocephalus, otoacoustic emissions, ventriculoperitoneal shunt

Abbreviations: ART = acoustic reflex threshold; CSF = cerebrospinal fluid; CA = cochlear aqueduct; DPOAE = distortion product otoacoustic emission; ICP = intracranial pressure; SNHL = sensorineural hearing loss; VP = ventriculoperitoneal

Unilateral Hearing Loss in Children with Shunt-Treated Hydrocephalus

Susan E. Spirakis*
Raymond M. Hurley†

* Audiology Department, Children’s Medical Service of Hillsborough County, Tampa, FL
† Department of Communication Sciences and Disorders, University of South Florida, Tampa, FL

Reprint requests: Raymond M. Hurley, Ph.D., Department of Communication Sciences and Disorders, University of South Florida, 4202 East Fowler Avenue—PCD 1017, Tampa, FL 33620-8150; Phone: 813-974-9784; Fax: 813-974-0822; E-mail: rhurley@chuma1.cas.usf.edu
Hydrocephalus results from an excessive accumulation of cerebrospinal fluid (CSF) in and around the brain due to an imbalance between CSF production and absorption, resulting in an increase in intracranial pressure (ICP) (Jackson, 1980; Pleasants, 1982). The ventricular system located within the cranium is responsible for CSF production and circulation. The CSF is produced in the choroid plexus, a highly specialized capillary bed located in the ventricles. CSF exits the ventricular system through the fourth ventricle to circulate through the spinal canal and subarachnoid spaces. Arachnoid villi within the subarachnoid space serve to filter the CSF into the venous system (Jackson, 1980; Pleasants, 1982; Marshall and Ross, 1984; Zemlin, 1998).

Hydrocephalus can be congenital or acquired resulting from genetic inheritance, spina bifida, meningitis, tumors, head injury or intraventricular hemorrhage. Hydrocephalus is classified as communicating or noncommunicating. In communicating hydrocephalus, CSF flows through the ventricular pathway unobstructed and into the subarachnoid space but is not absorbed into the venous circulation. Consequently the production of CSF exceeds its ability to be absorbed. The increased volume of fluid results in an increase in intracranial pressure. With noncommunicating hydrocephalus, an obstruction within the ventricular system impedes the CSF from adequately entering the subarachnoid space, resulting in increased CSF volume that distends the ventricles and increases ICP (Jackson, 1980; Pleasants, 1982; Marshall and Ross, 1984; Zemlin, 1998).

Standard medical treatment for hydrocephalus is the placement of a shunting device to help eliminate the excessive cerebrospinal fluid and to maintain a more normal intracranial pressure. Ventriculoperitoneal (VP) shunting is the preferred procedure as there is a lower incidence of serious complications. There are fewer infections, and fewer revisions, than those associated with other shunting procedures (Keucher and Meaby, 1979). The majority of VP shunts are placed in the patient's right side, as this is the nondominant cerebral hemisphere in the majority of patients. The neurosurgeon inserts a ventricular catheter into the right lateral ventricle. The catheter is coupled to a pressure valve and a reservoir that is housed within the patient's skull that in turn is coupled to a peritoneal catheter. This system of catheters allows for the excessive CSF to drain into the patient's peritoneal cavity where it is readily absorbed by the body (Pleasants, 1982; Marshall and Ross, 1984).

Ventriculoperitoneal Shunt and Hearing-Loss Mechanism

While the prevalence or risk of hearing loss in shunt-treated hydrocephalic children has not been established, the anatomical

Palabras Clave: Acueducto coclear, hipoacusia, hidrocefalo, emisiones otoacústicas, derivación ventriculo-peritoneal.

Abreviaturas: ART = umbral de reflejo acústico; CSF = líquido cefalo-raquídeo; CA = acueducto coclear; DPOAE = emisión otoacústica por producto de distorsión; ICP = presión intra-craneana; SNHL = hipoacusia sensorineural; VP = ventrículo-peritoneal

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relationship between the fluid spaces of the inner ear and the central nervous system allows the development of shunt-induced hearing loss (see Figure 1). Specifically, in the otic capsule, pressure homeostasis between the cochlear fluids and CSF is primarily maintained by two mechanisms (Durrant and Lovrinic, 1995). First, the cochlear aqueduct (CA), which allows direct communication between perilymph and CSF, controls perilymphatic pressure. Second, the endolymphatic sac, which lies within the subdural space surrounded by CSF, controls endolymphatic pressure. Although there is not direct communication between endolymph and CSF, the surrounding CSF pressure is easily transmitted to the endolymph sac (Tandon et al, 1973).

The CA would appear to be at the center of decrease in hearing after the loss of CSF. The CA located at the basal end of the cochlea is a bony canal filled with a mesh of loose connective tissue. It allows communication between the perilymph of the scala tympani and the CSF of the subarachnoid space in the posterior cranial fossa. Only small amounts of fluid are thought to move between the cochlea and the subdural space since only the membranous windows are compliant in the bony cochlear capsule (Walstead et al, 1994; Stoeckli and Bohmer, 1999). When CSF pressure changes occur, the transmitted variations give rise to very small displacements within the cochlea as there is only a minimal pressure difference between endolymph and perilymph. However, these pressure differentials can effect displacement of Reissner’s membrane and affect hearing. This link between hearing loss and low perilymphatic pressure has been confirmed using animal models. Funai et al (1988) reported threshold elevations after perilymphatic aspiration and bulging, and collapse and rupture of Reissner’s membrane. Arenberg and colleagues (1988) performed electrocochleography on guinea pigs following drainage of perilymph and demonstrated a change in the summating potential reflecting a change in position of the basilar membranes that was confirmed by histological studies.

**Reported Hearing Loss and Shunting**

A study by Lopponen et al (1989) reported a 38 percent prevalence of high-frequency sensorineural hearing loss (SNHL) within their sample of 47 shunt-treated hydrocephalic children. The authors defined hearing loss as a pure-tone average (500, 1000, and 2000 Hz) of greater than 15 dB. They defined a high-frequency hearing loss as a threshold of 20 dB or poorer at either 4000, 6000, or 8000 Hz. Their findings revealed that 18 children met their criteria for high-frequency hearing loss. Half of the children had bilateral high-frequency hearing loss while half demonstrated a unilateral loss. There was no indication in the unilateral loss cases whether the hearing loss ear was concurrent with the side of shunt placement. Of the 18 participants identified as having high-frequency SNHL, 11 cases were reported to have their hearing loss attributed to retrocochlear dysfunction. This determination was based on a contralateral acoustic reflex threshold (ART) of 100 dB or greater at 1000 Hz, an ART/HTL difference of greater than 60 dB, and the absence of middle-ear pathology. Lopponen et al (1989) hypothesized that long-term shunting may cause overdrainage of the CSF and produce cranial base hypoplasia resulting in brain stem involvement and elevated ARTs. However, their ART criterion for determining retrocochlear involvement is quite different from that traditionally used in clinical practice (Silman and Gelfand, 1981). Clearly, the Lopponen et al (1989) criterion would identify more children having retrocochlear dysfunction than would be identified using conventional ART norms.
There are several adult case studies describing hearing loss associated with hydrocephalus that is similar to the hearing loss experienced with endolymphatic hydrops. These studies have consistently demonstrated bilateral low-frequency SNHL that is resolved once the excessive ICP was relieved following shunt placement (Tandon et al, 1973; Barlas et al, 1983). Most recently, Stoeckli and Bohmer (1999) presented a case study of an adult who acquired hydrocephalus secondary to a subarachnoid hemorrhage following head trauma and was treated with the placement of a VP shunt. This patient experienced a persistent bilateral low-frequency hearing loss following shunt placement that was present one year postoperatively. Collectively, these articles may suggest that pressure variations in CSF can be transmitted to the labyrinth possibly by the cochlear aqueduct (CA), resulting in hearing loss (Tandon, et al 1973; Michel and Brusis, 1992; Walstead et al, 1994).

The literature that examines hearing loss following reduction of CFS and decreased ICP following neurosurgery, lumbar puncture, and spinal anesthesia in adults is relevant to the issue of shunt-treated hydrocephalus and hearing loss in children. Walstead et al (1994) looked specifically at the prevalence of hearing loss in adults following neurosurgery, which involved puncture or drainage of CSF from the subdural space. They reported a 53 percent prevalence of SNHL as defined as a shift of 15 dB or greater from presurgical thresholds that resolved within one week of surgery. The transient loss was most common at the low frequencies (125, 250, and 500 Hz) and at high frequencies (4000 and 8000 Hz). The authors hypothesized that the hearing loss results from a decrease in pressure or volume of the CSF with a concomitant reduction in perilymphatic fluid.

To date, there has been little research that explores the characteristics of hearing loss associated with shunt placement. Further, the literature that has been published is conflicting on the basic question of hearing loss etiology. Accordingly, this study investigated further the characteristics of hearing loss in VP-shunted hydrocephalic children. Since hydrocephalus can be congenital (approximately 1 in 500 births) or acquired, and is a lifelong chronic condition, identifying the characteristics of hearing loss in VP-shunted hydrocephalic children is important.

METHODS

Subjects

Twelve children participated in this study. Seven girls and five boys received VP shunts to treat hydrocephalus secondary to intraventricular hemorrhage or spina bifida. The children ranged in age from 7 years to 16 years, 6 months with a mean age of 12 years, 3 months. The criteria for inclusion in this study were: (1) a diagnosis of hydrocephalus; (2) the presence of a VP shunt; (3) the capability to perform audiometric testing; and (4) a signed parental consent form. The hydrocephalus was noncommunicating, and was either congenital or acquired. They received their shunts between the ages of 2 days and 15 months old. Eight of the children had right-sided shunts; three had left-sided shunts while one child had bilateral shunts, with the right shunt nonpatent and the left shunt patent. All of the other children’s shunts were patent and functional, as confirmed by neurological evaluations. The children were selected from the Children’s Medical Service of Hillsborough County active patient caseload based on their diagnostic codes. The children’s medical records were reviewed for inclusion and exclusion factors once parental consent was obtained.

Children were excluded from participation in the study if they had other major indicators for possible SNHL, that is, familial history, cerebral palsy, cytomeglovirus, congenital syphilis, or kernicterus. This determination was made by reviewing their medical records and by parental report. Children with active middle-ear pathology were also excluded from participation.

Procedures

Otoscopy was performed on each of the selected children. The children were assessed using a battery of audiometric test procedures that included pure-tone testing, immittance testing and distortion product otoacoustic emission (DPOAE) testing. Pure-tone audiometric testing was completed using a Beltone Model 110 audiometer or a Madison OB822 audiometer equipped with either TDH-39 or TDH-50 encased in MX/AR-41 cushions. The audiometers were recently
calibrated (American National Standards Institute, 1996), and testing was completed in a sound-treated test booth. Air-conduction thresholds were obtained at 500, 1000, 2000, 4000, 6000, and 8000 Hz using either a play audiometry paradigm or hand-raising response mode. Hearing loss was defined as a pure-tone threshold ≥ 20 dB HL at any test frequencies or a 15 dB difference between the two ears at 4000, 6000, and 8000 Hz.

Distortion product otoacoustic emissions (DPOAEs) were recorded (Grason-Stadler Model 60) at 812, 1000, 1280, 1590, 2030, 2560, 3180, and 4030 Hz using f1 = 65 dB SPL and f2 = 55 dB SPL with f1/f2 = 1.22. Tympanometry and ARTs were obtained (Grason-Stadler Model 33) with a 220 Hz probe tone. The ARTs were measured ipsilaterally at 1000 Hz, and contralaterally at 500, 1000, 2000, and 4000 Hz.

RESULTS

Pure-tone air-conduction thresholds were obtained at 500, 1000, 2000, 4000, 6000, and 8000 Hz bilaterally for all 12 participants. Of the 12 children, 10 (83%) demonstrated a high-frequency unilateral SNHL at 4000, 6000, and 8000 Hz in the ear ipsilateral to patent shunt placement. Eight of the children had patent right-sided VP shunts. Six demonstrated a significant right-ear high-frequency hearing loss. Three children had a patent left sided VP shunt. All three demonstrated a significant left-ear high-frequency hearing loss. One child had bilateral shunts, a nonpatent right shunt, and a patent left shunt. This child demonstrated a significant left-ear high-frequency hearing loss consistent with the side of the patent shunt. None of the children demonstrated a hearing loss in the nonshunt ear. A comparison of the mean pure-tone thresholds of the ear ipsilateral to the shunt (shunt ear) to those of the contralateral ear (nonshunt ear) is displayed in Figure 2. Standard error bars rather than the standard deviation are used to display the variability. Inspection of Figure 2 suggested that the threshold differences at 4000, 6000, and 8000 Hz might be significant. This was confirmed by the Wilcoxon matched-pairs statistic. These were significantly (p < .01) poorer thresholds for these frequencies in the shunt ear.

DPOAEs were obtained bilaterally on 9 of the 12 participants and were all at least 6 dB above the noise floor. Figure 3 graphically displays the mean OAE data and standard error bars. Inspection of Figure 3 suggests that a significant ear difference in DPOAE amplitude may exist at 2560, 3150, and 4036 Hz. The Wilcoxon matched-pairs statistic demonstrated a significantly (p < .05) reduced DPOAE amplitude at 2560 and 3150 Hz in the shunt ear. While the DPOAE amplitude was similarly reduced at 4036 Hz in the shunt ear, the reduction was not significant (p < .06).
Contralateral ARTs were obtained for all children at 500, 1000, 2000, and 4000 Hz bilaterally as were the ipsilateral ARTs at 1000 Hz. The mean 1000 Hz ipsilateral ARTs were 90 dB (±1SD = 10.62) and 86.36 dB (±1SD = 8.97) for the shunt ear and nonshunt ear, respectively. Figure 4 illustrates the ART data. The Wilcoxon matched-pairs sign rank test failed to reveal a significant difference between the ipsilateral and contralateral ARTs for the shunt ear and nonshunt ears at any test frequency.

In order to determine if the majority of ARTs were abnormal, a Binomial Test was applied. Designation of normal/abnormal for the contralateral ARTs was based on the data of Silman and Gelfand (1981) while the data of Wiley et al (1987) were used to classify the ipsilateral ARTs. The results of these analyses are presented in Table 1. Inspection reveals that a significant number of contralateral ARTs were abnormal at 500, 1000, and 4000 Hz but not at 2000 Hz. Conversely, a majority of ipsilateral ARTs at 1000 Hz were normal.

DISCUSSION

The difference in pure-tone thresholds between the patent shunt versus nonshunt ear was significant (p < .01). This unilateral finding is in contrast to the neurosurgical patients described by Walstead et al (1994) who experienced bilateral high-frequency hearing losses after lumbar puncture or drainage of CSF during surgery. However, the hearing loss resolved within one week postsurgery once the CSF level returned to normal. The authors hypothesized that the decrease in volume and pressure in the CSF transmitted to the perilymph by the CA resulted in bilateral endolymphatic hypertension, which produced the observed bilateral changes in hearing. In contrast, all of the children in the present study have noncommunicating hydrocephalus that requires a permanent unilateral VP shunt with active CSF drainage. The unilateral SNHL ipsilateral to shunt placement in the present in the study is consistent with the pathophysiology model for hearing loss following loss of CSF proposed by Walstead et al (1994); however, it is a unique finding due to the site of drainage and type of shunt used.

Similar to the children in the present study, the children in the Lopponen et al (1989) study had long-term shunt placement and high-frequency hearing losses. Unlike the children in the present study, who only demonstrated unilateral hearing losses, the participants in the Lopponen et al study (1989) showed an equal number of bilateral and unilateral high-frequency hearing losses. There is not a reference as to whether the unilateral hearing losses occurred in the ear ipsilateral to shunt placement; however, the authors did not report a significant difference between the occurrence of right-ear versus the left-ear unilateral losses. Recall that Lopponen et al (1989) attributed the observed hearing loss

### Table 1  Binomial Test Results for the Number of Significant Abnormal Acoustic Reflexes Thresholds (ARTs)

<table>
<thead>
<tr>
<th>Binomial Test</th>
<th>Acoustic Reflex (Hz)</th>
<th>500 Hz</th>
<th>1000 Hz</th>
<th>2000 Hz</th>
<th>4000 Hz</th>
<th>Ipsi 1000 Hz</th>
</tr>
</thead>
<tbody>
<tr>
<td>z value</td>
<td></td>
<td>3.84</td>
<td>2.14</td>
<td>1.63</td>
<td>2.86</td>
<td>-2.86</td>
</tr>
<tr>
<td>p value</td>
<td></td>
<td>&lt;.01</td>
<td>&lt;.05</td>
<td>&gt;.05</td>
<td>&lt;.05</td>
<td>&lt;.05</td>
</tr>
</tbody>
</table>
to a retrocochlear dysfunction in 11 of these children based on the difference between the ART and the pure-tone threshold. Their criterion for determining retrocochlear involvement utilized a smaller difference value between the ART and the pure-tone threshold than is traditionally used in clinical practice (Silman and Gelfand, 1981). This may have resulted in a greater number of children being identified with retrocochlear involvement. In the present study, there is not a significant difference between the ipsilateral and contralateral ARTs for the shunt and the nonshunt ears. However, there were a significant number of contralateral ARTs that exceeded 90th percentile cutoff value (Silman and Gelfand, 1981) while a significant number of ipsilateral ARTs at 1000 Hz was normal. Thus, the acoustic reflex results reflect a brain stem dysfunction pattern (Jerger and Jerger, 1975, 1977; Jerger et al, 1979). These findings support the hypothesis by Lopponen et al (1989) that long-term shunting may cause excessive drainage of CSF and produce cranial base hypoplasia resulting in brain stem involvement and elevated ARTs.

Although a patent CA is needed for the hearing loss to occur after loss of CSF (Walstead et al, 1994), a dilated CA has been implicated as a factor in SNHL after CSF drainage (Jackler and Hwang, 1993). The CA is considered to be patent in children and become gradually obstructed by fibrous material with age (Durrant and Lovrinic, 1995). Thus, variations in the age dependent patency of the CA may affect CSF pressure in the perilymphatic space and may have a differential effect on the two ears of the same person. In patients with a dilated CA, small changes in CSF pressure are easily transmitted and can produce cochlear dysfunction (Tandon et al, 1973). All of the participants of the present study have a permanent, unilateral, patent VP shunt that actively regulates and drains CSF. These children are demonstrating a unilateral high-frequency hearing loss in the ear ipsilateral to the patent shunt. DPOAE measurements demonstrate reduced emission amplitude in the patent shunt ear that is consistent with cochlear dysfunction (Tandon et al, 1973). None of the children demonstrated a hearing loss in the nonshunt ear. The acoustic reflex pattern indicates brain stem involvement similar to other brain stem dysfunctions (Jerger and Jerger, 1975, 1977). Accordingly, the data supports cochlear dysfunction in the ear ipsilateral to the patent shunt as the causative factor of the hearing loss with concomitant brain stem dysfunction.

In summary, the results of this study show that 83 percent of the children exhibited a SNHL high-frequency hearing loss in the shunt ear without hearing loss in the nonshunt ear. We hypothesize that the pathophysiology of the hearing loss is related to a patent CA combined with a patent VP shunt that provides an open communication pathway for fluid pressure variations. The patent VP shunt is actively draining excessive CSF. As the shunt reduces the volume of CSF, and there is a decrease in ICP, a concomitant decrease in perilymphatic fluid pressure occurs due to the open pathway to the perilymph provided by a patent CA. This decrease in perilymphatic fluid, although minimal, may be sufficient to disrupt cochlear hydrodynamics and has a deleterious effect on hearing in the ear ipsilateral to shunt placement. We further suggest that the hypertensive state of the endolymphatic pressure within the membranous labyrinth as a result of the decreased perilymph pressure affects the displacement properties of Reissner’s membrane leading to cochlear dysfunction and hearing loss. We suggest that the hearing loss is in the high-frequency region due to the anatomical location of the CA in the otic capsule; the CA being located in the basal portion of the cochlea. The audiometric threshold findings, and the reduced DPOAE's in the shunt ear, suggest that the hearing loss is cochlear (sensory) in etiology and permanent in nature as long as the shunt remains patent. We suggest that the auditory brain stem involvement as evidenced by the ART pattern is the result of the active drainage of CSF from the subdural space by a patent shunting mechanism. As hypothesized by Lopponen et al (1989), the long-term shunting may result in anatomical abnormalities of the skull-base resulting in disturbance to the brain stem. Two children failed to exhibit any hearing loss. Review of their ART pattern also failed to demonstrate a brain stem pattern. The absence of both hearing loss and an ART auditory brain stem pattern may reflect a better homeostatic relationship between the cochlear fluids and CSF. We hypothesize that their CA may not be patent, or they are not experiencing

Acknowledgment. This was a professional research project (PRP) completed in partial fulfillment for the degree of Doctor of Audiology at the University of South Florida.

REFERENCES


