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Incidence of unilateral, high frequency, sensorineural hearing loss in shunt treated hydrocephalic children ipsilateral to shunt placement

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Incidence of Unilateral, High Frequency, Sensorineural Hearing Loss in Shunt Treated Hydrocephalic Children Ipsilateral to Shunt Placement

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Professional Research Project
Submitted to the Faculty of the University of South Florida
In partial fulfillment of the requirements for the degree of

Doctor of Audiology

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(ABSTRACT)

The purpose of this study was to investigate further the characteristics of hearing loss in ventriculoperitoneal (VP) shunted hydrocephalic children. Twelve VP shunt treated hydrocephalus children participated in this study. The etiology of the hydrocephalus was either intraventricular hemorrhage or spina bifida. A recent neurological examination reported the shunt to be patent in each child. Audiometric examination included pure tone air conduction thresholds, tympanometry, contralateral and ipsilateral acoustic reflex thresholds (ARTs), and distortion product otoacoustic emissions (DPOAE’s). 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 findings coupled with the decrease in DPOAE amplitude in the shunt ear, the hearing loss appears to cochlear in nature. It is hypothesized that cochlear hydrodynamics are disrupted as the result of fluid pressure reduction within the perilymph being transmitted via a patent cochlear aqueduct as a reaction to the reduction of CSF via a patent shunt. 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.
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INTRODUCTION

Hydrocephalus results from an excessive accumulation of cerebrospinal fluid (CSF) in and around the brain resulting in an increase in intracranial pressure (ICP), (Jackson, 1980). The ventricular system located within the cranium is responsible for CSF production and circulation. The CSF is produced in the choroid plexus that is a highly specialized capillary bed located in the ventricles. The ventricles are all connected by specific structures to allow for CSF circulation. The fourth ventricle has two openings, the foramen of Sylvius and the foramen of Luschka, which allow CSF to circulate and bathe the spinal canal and subarachnoid spaces. Arachnoid villi within the subarachnoid space serve to filter the CSF into the venous system (Jackson, 1980; Zemlin, 1998).

Hydrocephalus may be classified as communicating or non-communicating. In communicating hydrocephalus, CSF is circulated through the ventricular system 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 non-communicating hydrocephalus, an obstruction within the ventricular system impedes the CSF from adequately entering the subarachnoid space. The obstruction may be due to a tumor, a hemorrhage or improper development of CSF in the circulatory pathway. With non-communicating hydrocephalus, increased CSF volume distends the ventricles and increases ICP (Jackson, 1980; Pleasants, 1982; 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, i.e. fewer infections, fewer revisions, than associated with other shunting procedures (Keucher & Meaby, 1979). The majority of VP shunts are placed in the patient’s right side, as this is the non-dominant cerebral hemisphere. The neurosurgeon surgically 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 (Marshall & Ross, 1984; Pleasants, 1982).

Located in the petrous portion of the temporal bone is the inner ear that has an intricate system of fluid filled spaces that are connected to the fluid filled spaces of the central nervous system. Mechanisms exist to maintain the homeostasis of pressure between the two cochlear fluids (perilymph and endolymph) and between the cochlear fluids and the CSF (Durrant & Lovrinic, 1984). Perilymphatic pressure is maintained via the cochlear aqueduct (CA) which allows direct communication between perilymph and CSF. Endolymphatic pressure is maintained by the endolymphatic sac that lies within the subdural space and is surrounded by CSF. Although there is not direct communication between endolymph and CSF, the surrounding CSF pressure is easily transmitted to endolymph (Tandon, Sinha, Kacker, Saxena & Singh, 1973).

The neurological manifestations and sequel associated with untreated hydrocephalus and with VP shunt placement in children and adults is well-documented (Boynton, Boynton, Merit, Voucher, James & Bear, 1986; In, Oh, Brown & Steers, 1992; Servo, Ferrule, Heikkinen, Andiron & von Wendt, 1990). However, there is a dearth of data regarding the incidence or risks of hearing loss in shunt treated hydrocephalic children. The singular study by Lopponen, Sorri, Serlo and von Wendt (1989) reported a 38% 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, and a high frequency hearing loss as a threshold of 20 dB or worse at 4000, 6000 or 8000 Hz. Results of their pure tone 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. It was not mentioned if the hearing loss present in the cases of unilateral loss was concurrent with the side of shunt placement. There was not, however, an ear effect associated with the unilateral hearing loss. Of the 18 participants identified as having high frequency hearing loss, 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/air conduction threshold difference of greater than 60 dB, and the absence of middle ear pathology. Loppenen et al. (1989) hypothesized that long-term shunting may cause over-drainage 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 than that traditionally used in clinical practice (Silman & Gelfand, 1981). Clearly, their criterion would identify more children as having retrocochlear dysfunction than would be identified using conventional ART norms.

Relevant to the issue of shunt-treated hydrocephalus and hearing loss in children, is the literature that examines hearing loss following reduction of CFS and decrease ICP following neurosurgery, lumbar puncture, and spinal anesthesia in adults. These articles suggest that pressure variations in CSF can be transmitted to the labyrinth by the cochlear aqueduct (CA), to the inner ear by the endolymphatic duct and/or the perilymphatic duct (Michel & Bruis, 1992; Tandon, et al., 1973; Walstead, Nielsen & Borum, 1994).

Walstead et al. (1994) looked specifically at the incidence of hearing loss in adults following neurosurgery, which involved puncture or drainage of CSF from the subdural space. They reported a 53% prevalence of SNHL as defined as a shift of 15 dB or greater from pre-surgical 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 the hearing loss results from a decrease in pressure and/or volume of the CFS with a concomitant reduction in perilymphatic fluid.

There are several adult case studies describing hearing loss due to hydrocephalus. Various shunting procedures, including VP shunting, were used to relieve ICP. These studies have consistently demonstrated bilateral low frequency hearing loss which is usually transient in nature that resolves following shunt placement (Barlas, Gokay, Turantan & Baserer, 1983; Tandon, et al., 1973).
Stockeli and Bohmer (1999) presented a case study of an adult who acquired hydrocephalus secondary to a subarachnoid hemorrhage following head trauma. The hydrocephalus 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 post-operatively. The authors of the study concluded that persistent hearing loss in patients who have undergone shunt placement for hydrocephalus is perhaps an underestimated complication.

To date, there has been little research that explores the characteristics of hearing loss that maybe associated with shunt placement. Further, the literature that has been published is conflicting on the basic question of hearing loss etiology. Accordingly, it is the purpose of this study to investigate further the characteristics of hearing loss in VP shunted hydrocephalic children. As the incidence of hydrocephalus at birth is approximately 1-3 per 1000 births and children can experience postnatal hydrocephalus, identifying the characteristics of hearing loss in VP shunted hydrocephalic children has merit.

**METHODS**

**Subjects**

Participants in this study consisted of twelve children, seven girls and five boys, who 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) diagnosed hydrocephalus; 2) the presence of a VP shunt; 3) capable of performing audiometric testing; and 4) a signed parental consent form. The hydrocephalus was non-communicating, and was either congenital or acquired. They received their shunts between the age of 2 days to 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 non-patent and the left shunt patent. All of the other children’s shunts were patent and functional 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 sensorineural hearing loss (SNHL) i.e., 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 and Instrumentation

Otoscropy was performed on each of the selected subjects. The subjects 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 audiometry testing was completed using a Beltone Model 10 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 (ANSI, 1996) and testing was completed in a calibrated sound treated booth. Air conduction thresholds were obtained at 500, 1000, 2000, 4000, 6000 and 8000 Hz using a modified method of limits incorporated into either a play audiometry paradigm or hand-raising response mode. This study’s definition of a hearing loss is a pure tone threshold $\geq 20$ dB at any test frequencies; or a 15 dB difference between the two ears at 4000, 6000 and/or 8000 Hz.

Distortion product otoacoustic emission (DPOAE) testing was completed using the Grason-Stadler DPOAE 60 equipment that was calibrated in accordance with manufacturer’s specifications. The equipment was also calibrated daily using its calibration cavity. DPOAEs were recorded at 812, 1000, 1280, 1590, 2030, 2560, 3180, and 4030 Hz using $f_1 = 65$ dB SPL and $f_2 = 55$ dB SPL with $f_1/f_2 = 1.22$. This testing was also completed within a calibrated sound booth.

Tympanometry and ARTs were obtained using a calibrated (ANSI, 87) Grason-Stadler 33 immittance system. A 220 Hz probe tone was used measure the tympanograms and to monitor the ARTs that were measured ipsilaterally at 1000 2000 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 twelve participants. Of the twelve subjects, ten (83%) of them demonstrated a high frequency sensorineural (SNHL) unilateral hearing loss at 4000, 6000 and 8000 Hz in the ear ipsilateral to patent shunt placement. Eight of the children had patent right-sided VP shunts with six of them demonstrating a significant right ear high frequency hearing loss. Three children had a patent left sided VP shunt with all of the demonstrating a significant left ear high frequency hearing loss. One child had bilateral shunts, a non-patent 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 subjects demonstrated a hearing loss in the non-shunt ear. Table 1 reports the means and standard deviations for these pure tone data. A comparison of the mean pure tone thresholds of the ear ipsilateral to the shunt (shunt ear) to those of the contralateral ear (non-shunt ear) is displayed in Figure 1. Standard error bars rather than the standard deviation are used to display the variability in Figure 1. Standard error bars are utilized in Figures 1–4 for better representation of variability and to more graphically depict the obvious differences between the results obtained from the shunt versus the non-shunt ear. Inspection of Table 1 and Figure 1 suggests that the threshold differences at 4000, 6000, and 8000 Hz may be significant. This was confirmed by the Wilcoxon matched-pairs statistic that demonstrated significantly (p<. 01) poorer thresholds for these frequencies in the shunt ear.

Table 1. Mean and standard deviation (±1SD) air conduction thresholds in dB HL for the non-shunt and shunt ears of the 12 subjects.

<table>
<thead>
<tr>
<th>Ear</th>
<th>Hearing Threshold Level in dB</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>500 Hz</td>
</tr>
<tr>
<td>Non-Shunt</td>
<td>15.00 (5.64)</td>
</tr>
<tr>
<td>Shunt</td>
<td>13.33 (5.77)</td>
</tr>
</tbody>
</table>
DPOAE’s were obtained bilaterally on nine of the twelve participants. Table 2 reports the means and standard deviations for these DPOAE data. A graphic display of these data appears in Figure 2, but uses standard error bars rather than the standard deviation to illustrate variability. Inspection of Figure 2 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 Hz in the shunt ear. While the DPOAE amplitude was similarly reduced at 3150 and 4036 Hz in the shunt ear, the reduction was not significant being p< .08 and p< .09, respectively.

**Table 2.** Mean and standard deviation (±1SD) distortion product otoacoustic emission (DPOAE) amplitude in dB SPL for the non-shunt and shunt ears of the 12 subjects.

<table>
<thead>
<tr>
<th>Ear</th>
<th>2 f₁-f₂ Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>812   1000 1280 1590 2030 2560 3180 4030</td>
</tr>
<tr>
<td>Non-Shunt</td>
<td>6.67  9.00 9.56 5.89 4.11 6.00 6.78 3.78</td>
</tr>
</tbody>
</table>
Contralateral ART’s were obtained for all subjects at 500, 1000, 2000 and 4000 Hz bilaterally; however, only the ipsilateral ART at 1000 Hz was consistently measured. The mean 1000 Hz ipsilateral ARTs were 90 dB (±1SD = 10.62) and 86.36 dB (±1SD = 8.97) for the shunt ear and non-shunt ear, respectively. Table 3 reports the means and standard deviations for the contralateral ART data. Figure 3 illustrates the ART data, but utilizes the standard error bars rather than the standard deviation to depict variance. The Wilcoxon matched-pairs sign rank test failed to reveal a significant difference between the ipsilateral and contralateral ARTs for the two ears at any of the test frequencies.

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 & Gelfand (1981) while the data of Wiley, Oviatt and Block (1987) was used to classify
the ipsilateral ARTs. The results of these analyses are presented in Table 4. Inspection of Table 4 reveals that a significant number of contralateral ARTs were abnormal at 500, 1000 Hz and 4000 Hz, but not at 2000 Hz. Conversely, a significant majority of ipsilateral ARTs at 1000 Hz were normal.

**Table 3.** Mean and standard deviation (±1SD) contralateral acoustic reflex thresholds (ARTs) in dB HL for the non-shunt and shunt ears of the 12 subjects.

<table>
<thead>
<tr>
<th>Ear</th>
<th>Acoustic Reflex Thresholds</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>500 Hz</td>
</tr>
<tr>
<td>Non-Shunt</td>
<td>102.27</td>
</tr>
<tr>
<td></td>
<td>(7.86)</td>
</tr>
<tr>
<td>Shunt</td>
<td>102.27</td>
</tr>
<tr>
<td></td>
<td>(10.09)</td>
</tr>
</tbody>
</table>

**Table 4.** 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>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>500 Hz</td>
</tr>
<tr>
<td>z value</td>
<td>3.84</td>
</tr>
<tr>
<td>p. value</td>
<td>&lt;.01</td>
</tr>
</tbody>
</table>
DISCUSSION

The difference in pure tone thresholds between the patent shunt versus non-shunt ear was significant ($p < 0.01$). This unilateral finding is in contrast to the neurosurgical patients described by Walstead et al. (1994) who experienced bilateral high frequency hearing losses after puncture or drainage of CSF during surgery. A permanent shunt with an active pressure-regulating pump was not used with these patients. In the Walstead et al. (1994) subjects, the loss of CSF was temporary and resolved within one-week post-surgery. The authors stated that the decrease in volume and pressure in the CSF transmitted to the perilymph by the CA resulted in endolymphatic hypertension, which produced the observed changes in hearing.

Similar to the subjects in the present study, the children in the Loppenen et al. (1989) study had long-term shunt placement and high frequency hearing losses. Unlike the children in this study who only demonstrated unilateral hearing losses, the participants in the Loppenen et al. study (1989) demonstrated 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 Loppenen et al. (1989) reported that 18 of their 47 subjects demonstrated high frequency hearing losses. Loppenen et al. (1989) attributed the observed hearing loss 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 & 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 non-shunt ears. However, there were a significant number of contralateral ARTs that exceeded 90th percentile cutoff value (Silman & Gelfand, 1981) while a significant number of ipsilateral ARTs at 1000 Hz was normal. Thus, the acoustic reflex results reflect the classic brain stem dysfunction pattern (Jerger & Jerger, 1975; Jerger & Jerger, 1977; Jerger, Jerger & Hall, 1979). These findings support the hypothesis by Loppenen, 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 ART’s.

Knowledge of the anatomy and physiology of the CA is relevant to understanding the theories of decrease in hearing after the loss of CSF. The CA, which is 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, as the cochlea is a hard-walled bony capsule in which only the membranous windows are compliant (Stoeckli & Bohmer, 1999). In cases of CSF pressure changes, the transmitted variations give rise to very small displacements within the cochlea because of the minimal pressure difference between endolymph and perilymph. However, these pressure differentials can effect displacement of Reissner’s Membrane and affect hearing. A perilymphatic hypotensive state may be created leading to relative endolymphatic hypertension, a situation similar to endolymphatic hydrops (Walstead et al., 1994). The connection between hearing loss and low perilymphatic pressure has been confirmed in some experimental studies. Funai, Hara and Nomura (1988) reported threshold elevations after perilymphatic aspiration. Their study also noted structural changes of Reissner’s membrane including bulging, collapse
and rupture. A study by Arenberg, Ackley, Ferraro and Muchnick (1988) performed electrocochleography on guinea pigs following drainage of perilymph and demonstrated a change in the summating potential indicating a change in position of the basilar membranes which was confirmed by histological studies.

A patent CA is needed for the hearing loss to occur after loss of CSF (Walstead, et al., 1994) while a dilated CA has been implicated as a factor in SNHL after CSF drainage (Jackler & Hwang, 1993). The CA is considered to be patent in children and become gradually obstructed by fibrous material with age (Durrant & Lovrinic, 1995). Thus, variations in the patency of the CA may affect the transmission of CSF pressure to the perilymphatic space depending on age and may have a differential effect on the two ears of the same person. In patients with a dilated CA, the CSF pressure would be transmitted easily, leading to cochlear dysfunction (Tandon et al., 1973). What is unique to the participants of this study is that they 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 non-shunt ear. The acoustic reflex pattern indicated brain stem involvement and similar to other brain stem dysfunctions, a normal audiogram can be obtained as seen in the co-existing normal hearing in the non-shunt ear (Jerger & Jerger, 1975; Jerger & Jerger, 1977). Accordingly, the data supports cochlear dysfunction in the ear ipsilateral to the patent shunt as the causative factor of the hearing loss with a concomitant brain stem dysfunction.

In summary, the results of this study demonstrate that 83% of the children are exhibiting a SNHL high frequency hearing loss in the shunt ear without hearing loss in the non-shunt ear. It is hypothesized that the pathophysiology of the hearing loss is related to a patent CA combined with a patent VP shunt which 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, is sufficient to disrupt cochlear
hydrodynamics and has a deleterious effect on hearing in the ear ipsilateral to shunt placement. It is further hypothesized 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. It is believed that the hearing loss is high frequency due to the anatomical location of the CA in the otic capsule; the CA being located in the basal portion of the cochlea. Based on the pure tone findings, and the reduced DPOAE’s in the shunt ear, it is believed that the hearing loss is cochlear (sensory) in etiology and permanent in nature as long as the shunt remains patent. It is believed that the brain stem dysfunction 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 Loppenen, et al., (1989) the long term shunting may result in anatomical abnormalities of the skullbase, causing brain stem disturbances. There were two children who failed to exhibit any hearing loss as defined by the study. Review of their ART pattern also failed to demonstrate a brain stem dysfunction. Accordingly, it is felt that the absence of hearing loss and brain stem dysfunction may reflect a better homeostatic relationship between the cochlear fluids and CSF. It is hypothesized that their CA may not be patent and/or they are not experiencing over-drainage of CSF as described by Huggare, Kantomaa, Ronning & Serlo, (1986) or Lopponen et al. (1989).

The current study is limited in scope due the small sample size of participants and the limited test battery to assess cochlear and retrocochlear function. Accordingly, future research into the prevalence and pathophysiology of hearing loss specific to shunt treated hydrocephalic children should include a larger number of subjects as well as radiographic studies of the participant’s CA’s. In addition, more detailed auditory testing should included microstructure audiograms, detailed DPOAE studies, auditory brain stem testing and electrocochleography.
REFERENCES


