HISTOPATHOLOGY OF THE STRIA VASCULARIS IN CHILDREN
Collin S. Karmody M.D., F.R.C.S.E.¹
Edgar Bachor M.D., Ph.D.²

¹ Department of Otolaryngology, Tufts University School of Medicine and the New England Medical Center
Boston, Massachusetts, USA
² Department of Otorhinolaryngology, SRH Zentralklinikum Suhl gGmbH, Germany

INTRODUCTION
Hearing loss in children, whether congenital or identified later, continues to be a challenging problem for parents and physicians. Unfortunately, this emotional entity generally remains somewhat of a puzzle because of a dearth of the most basic histopathological studies of the inner ear. Atrophy of the stria vascularis, presumably insidious, is regarded as a common cause of hearing loss in the elderly.¹ This stimulated our interest as to whether or not there were similar changes in the stria vascularis of newborns, infants and children that might shed some light on the pathophysiology of hearing loss in childhood.

MATERIAL AND METHODS
We examined 120 temporal bones from 71 infants between the ages of newborn to ten years by light microscopy from the temporal bone collection at Tufts University School of Medicine and the New England Medical Center. This collection of temporal bones is unique in that the specimens were unselected, and are, therefore, representative of the broad spectrum of otopathology in the general population. They were collected as they became available at autopsy, and were not procured from patients with a personal or family history of otologic problems. The causes of death were many and varied. In the pediatric group, the largest segment died from infections. All temporal bones were fixed in 10% formalin, decalcified and processed in the usual manner by the celloidin technique.¹ The specimens were sectioned in the horizontal plane at a thickness of 20 µm, every tenth section was stained with hematoxylin-eosin, and studied by light microscopy.

The stria was considered to be normal if all three layers (marginal, intermediate and basal) could be clearly identified and the cellular structure was normal at the light-microscopic level. Pathology of the stria vascularis was classified as
Histopathology of the Stria Vascularis in Children, Continued from page 1

(1) atrophy (2) widening (cystic change, infiltration or basophilic deposits) (3) anomalies (vascular or dysplasia) and (4) other (hyperpigmentation).

The stria was arbitrarily divided into three zones - upper, middle, and lower, according to the method of Zaytoun, the upper third being the area close to the insertion of Reissner’s membrane and the lower third adjacent to the spiral prominence (Fig. 1). Autolytic changes were determined by comparing the preservation of the associated organ of Corti and supporting cells. If there were no significant changes in the organ of Corti, then small interstitial spaces were classified as edema. Severe disintegration of the organ of Corti and similar changes, especially of the marginal layer and elevation of the stria from the spiral ligament indicated autolysis. If the layers of the stria vascularis were loose with substrial spaces, this was classified as degeneration, the end stage of which was necrosis. Atrophy was defined as cellular loss and reduction in size and shape in one of the three zones. In addition, we looked for abnormal cell types in the different layers. Vascular anomalies were defined as the presence of vessels that were abnormal in size, shape or number. In the pediatric population, it is frequently difficult, if not impossible, to distinguish between dysplasia and atrophy of the stria. We believe that when there is obvious maldevelopment of the inner ear, an aberration of the stria should be considered as a dysplasia. On the other hand, atrophy should be classified as a phenomenon that affects a normally developed organ.

RESULTS

The stria vascularis was normal in 75 (62.5%) specimens (Table 1). The next most frequent finding was atrophy in 31 ears (25.8%) (Fig. 2A). The most commonly affected area was the intermediate layer in the lower third (close to the spiral prominence). Narrowing was seen mainly in the upper third in older children, whereas in newborns, narrowing was primarily in the lower thirds—presumably a result of hypoplasia.

The stria vascularis was widened by cellular swelling, cellular hyperplasia or deposition of non-cellular material in 37 (30.8%), of which cyst formation was seen in 14 (11.6%) specimens (Fig. 2B). Macro-cysts were large fluid-filled spaces in the marginal or intermediate layer.

Deposits of basophilic non-cellular material were found mainly between the intermediate and the basal cell layers in 13 (10.8%) of temporal bones (Fig. 2D). Six of these patients had renal disorders such as nephrosclerosis or Potter’s syndrome. The other patients had died from non-renal problems. Infiltration with abnormal cells occurred in 10 (8.3%). In 3 children with leukemia, there was hemorrhage into the intermediate cell layer. In one patient with Niemann-Pick disease type A, the stria was loose and contained many plump cells with vacuolated cytoplasm which are typical of the disease.

Figure 1
A. Normal stria vascularis of the basal turn in a newborn infant.
B. Schematic representation of location of strial pathology used in this study.
Anomalies of the stria vascularis were identified in 15 (12.5 %) specimens (Fig. 2C). Vascular anomalies were found in 9 (7.5%) (Fig. 2E). Some vessels could be traced from their origin to their destination in the spiral ligament without formation of an intermediate capillary bed.

DISCUSSION

The most frequent pathological picture seen in our study was atrophy, which seemed to be related to infections. Johnsson and Hawkins, however, by comparing the temporal bones of young and elderly patients, found a gradual loss of capillaries in the spiral ligament with aging\(^4\). They postulated that the stria was essential for the development of the organ of Corti and even suggested that strial dysfunction preceded degeneration of the organ of Corti. Also, Fujimura et al\(^5\) found deposition of IgG and thickening of strial vessels beginning as early as the first week of life in dominant white spotting W/W\(\text{v}\) mice which are known to have hereditary sensorineural hearing loss.

Subotic et al\(^6\) described three types of changes of the stria vascularis in patients with documented congenital hearing loss: pseudocyst formation with dark-stained deposits, accumulation of endothelial cells or irregularly shaped cells in the basal coil of the cochlea, and atrophy of the stria vascularis in all turns of the cochlea.

The gene KCNQ1, known to be a potassium channel regulator, is present in the stria vascularis. Rivas and Francis in a study of congenitally deaf KCNQ1 knockout mice found marked atrophy of the stria, contraction of the endolymphatic compartment, collapse and adhesion of Reissner’s membrane and complete degeneration of the organ of Corti and the spiral ganglion\(^7\). They postulated that the histopathologic picture was comparable to that reported in humans with the Jervell and Lange-Nielsen syndrome. This study strongly suggests that the physiological integrity of the stria, possibly even in-utero, is essential for the physical development of the organ of Corti. Strial atrophy has been reported with many hearing problems, including presbycusis\(^1\), viral infections, ototoxicity, noise exposure etc. With viral infections, we frequently saw atrophy or cyst formations in the stria vascularis with specific

See Histopathology of the Stria Vascularis in Children, page 4

**Figure 2**

A. Atrophy of the stria in the middle third with accumulation of melanin in the upper and lower thirds.

B. Widening due to large cystic space in the intermediate layer of the stria.

C. Dysplasia of the upper third of the stria in a 3-day old girl.

D. Basophilic deposits between the intermediate and basal cell layer in a 18-month old boy with micropolygyria, hydrocephalus and glomerulosclerosis.

E. Vascular abnormality: there are several abnormal blood vessels in the intermediate cell layer of this 5-week girl with congenital microcephaly.
the changes that seemed to be related to the type of virus\(^8\). A granulomatous-like stria was previously reported in mumps\(^9\) and measles\(^10\), while in-utero infections with the cytomegalovirus\(^11\) or rubella virus\(^12\) showed strial hyperplasia and/or cystic degeneration.

Although in this and other studies, the tendency is to focus on the more generalized changes affecting the whole cochlea, it must be remembered that local mechanisms have been shown experimentally to control local homeostasis of inner ear fluids\(^13,14\) which alters function of local receptors. Localized cystic and other lesions may be asymptomatic or present with mild hearing loss.

Another important group of strial changes was the presence of basophilic deposits. Currently, the biochemical composition and etiology of these deposits is unknown. Zaytoun\(^2\) found basophilic deposits in 2% of temporal bones, which he related to patients with renal failure and suggested that immune complexes are a major source of these deposits. However, less than half of our group with basophilic deposits had renal disease, and infants with multiple anomalies had no more deposits than other groups.

In our cohort, widening of the stria vascularis was only a descriptive entity related to a variety of etiologies. Thickened strial vessels were found in one patient, but no detailed history was available. Similar thickening of the strial vessels have been described in patients with Type I diabetes\(^15\). Widening of the stria can also be caused by other phenomena such as infiltration with leukemic cells\(^16\), macrophages filled with lipids\(^3\), and hemorrhage due to leukemia\(^17\).

This study was confined to the stria vascularis. Although the stria impacts directly on the functioning of the inner ear, it also works in concert with the cells of the spiral ligament, Hensen’s cells and hair cells to achieve normal functioning of the scala media. The histopathology of this structure, therefore, must be integrated with the status of the rest of the scala media. Our premise is that the (pathologic) changes and lesions of the stria vascularis that we documented, although not correlated with detailed hearing tests, could provide the basis for some degree of hearing losses.

| Classification and frequency of histopathological findings in the human stria vascularis |
|-----------------------------------------------|------------------|
| n=120 temporal bones from 71 patients. 20 temporal bones showed more than one abnormality |

<table>
<thead>
<tr>
<th></th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>75</td>
</tr>
<tr>
<td>Atrophy</td>
<td></td>
</tr>
<tr>
<td>uniform</td>
<td>23</td>
</tr>
<tr>
<td>granulomas</td>
<td>8</td>
</tr>
<tr>
<td>Widening</td>
<td></td>
</tr>
<tr>
<td>cystic changes</td>
<td>14</td>
</tr>
<tr>
<td>infiltration</td>
<td>10</td>
</tr>
<tr>
<td>deposits (basophil)</td>
<td>13</td>
</tr>
<tr>
<td>Anomalies</td>
<td>9</td>
</tr>
<tr>
<td>vascular abnormalities</td>
<td>7.5%</td>
</tr>
<tr>
<td>dysplasia</td>
<td>6</td>
</tr>
<tr>
<td>Other</td>
<td>19</td>
</tr>
<tr>
<td>hyperpigmentation</td>
<td>10.7%</td>
</tr>
</tbody>
</table>
ACKNOWLEDGEMENTS
The authors would like to thank Julie A. Swan for her kind assistance in this study and Claudia Wacker, Medical photographer.

REFERENCES

Address for correspondence:
Collin S. Karmody, M.D.
Department of Otolaryngology
Tufts-New England Medical Center
750 Washington Street
Boston, MA 02111
e-mail: ckarmody@tufts-nemc.org

Otopathology Mini-Travel Fellowship Program
The NIDCD National Temporal Bone Registry is pleased to announce the availability of mini-travel fellowships. The fellowships provide travel funds for research technicians and young investigators to visit a temporal bone laboratory for a brief educational visit, lasting approximately one week. The emphasis is on the training of research assistants, technicians and junior faculty. The fellowships are available to:
1) U.S. hospital departments who aspire to start a new temporal bone laboratory 2) Inactive U.S. temporal bone laboratories that wish to reactivate their collections or 3) Active U.S. temporal bone laboratories that wish to learn new research techniques
Two fellowship awards will be made each year ($1,000 per fellowship). The funds may be used to defray travel and lodging expenses. Applications will be decided on merit.
Interested applicants should submit the following:
1) A 1-2 page outline of the educational or training aspect of the proposed fellowship 2) Applicant’s curriculum vitae 3) Letter of support from applicant’s temporal bone laboratory director or department chairman 4) Letter from the host temporal bone laboratory, indicating willingness to receive the traveling fellow

Applications should be sent to:
Saumil N. Merchant, M.D.
NIDCD National Temporal Bone Registry
Massachusetts Eye and Ear Infirmary
243 Charles Street
Boston, MA 02114
Scientific study of the human temporal bone and related brain tissue is a time-consuming process performed in highly specialized otopathology laboratories by researchers who are dedicated to enhancing our understanding of the pathology underlying disorders of hearing and balance. “Laboratory Spotlight” is a continuing series of articles offering a glimpse inside the laboratories in the United States and abroad conducting temporal bone research.

Temporal Bone Collection
Department of Otolaryngology
Tufts University School of Medicine
Tufts-New England Medical Center, Boston, Massachusetts, USA

The Temporal Bone Collection at the Department of Otolaryngology, Tufts University School of Medicine in Boston consists of over 200 human temporal bones. Of these, 120 bones are from children. The collection was started in 1970 by Dr. Collin Karmody, Professor of Otolaryngology at Tufts University School of Medicine. Most of the temporal bone specimens were processed using the standard celloidin technique, but a few were embedded in plastic.

This collection of temporal bones is unique in that the specimens were collected as they became available at autopsy; patients with a personal or family history of otologic disorders were not pre-selected. Therefore, the collection is representative of the broad spectrum of otopathology in the general population. The collection is also unique in that over half of the specimens are from children. In addition to human specimens, the collection also contains over 100 animal specimens, including tissue from squirrel monkeys, guinea pigs etc. as a result of various experiments performed over the years.

The laboratory stopped actively processing new specimens in 1994. It now serves as a repository of material collected and processed over the years. Dr. Collin Karmody continues his stewardship as custodian of the collection (see photo). The material continues to be in active use for scientific study, with ongoing collaborations with various faculty both within and outside the department of Otolaryngology. There has been a longstanding and particularly fruitful collaboration with Dr. Edgar Bachor, Chief of Otolaryngology at SRH Zentralklinikum Suhl gGmbH in Germany (see photo).
Scientific publications in otopathology from the Tufts-New England Medical Center collection:


PLEASE! Notify us of your change of address before you move.
Each undelivered newsletter is returned to the Registry office at
a cost of $.70. Our loss is over $1.00 per unit.
Thank you!

Free Brochures for your Office or Clinic about Temporal Bone Research and Donation

<table>
<thead>
<tr>
<th>That Others May Hear</th>
<th>The Gift of Hearing and Balance: Learning about Temporal Bone Donation</th>
</tr>
</thead>
<tbody>
<tr>
<td>is a short brochure</td>
<td>is a 16-page, full-color booklet which describes in more detail the benefits of temporal bone research. It also answers commonly asked questions regarding the temporal bone donation process.</td>
</tr>
<tr>
<td>which describes briefly the functions of the Registry, and answers commonly asked questions regarding the temporal bone donation process. (Dimensions: 9” x 4”)</td>
<td>(Dimensions: 7” x 10”)</td>
</tr>
</tbody>
</table>

If you are willing to display either or both of these brochures, please complete the form below and return it to the Registry by mail or fax. The brochures will be sent to you free of charge. Please circle the amount requested for each brochure or write in amount not listed.

That Others May Hear _____ 25  50  100
Enrollment Packets _____ 25  50  100
The Gift of Hearing and Balance: Learning about Temporal Bone Donation _____ 25  50  100
Newsletters _____ 25  50  100

NAME: ____________________________________________

ADDRESS: ________________________________________

ADDRESS: ________________________________________

CITY, STATE, ZIP: ________________________________________

TELEPHONE: ________________________________________

Mail or fax this form to the Registry at: NIDCD National Temporal Bone, Hearing and Balance Pathology Resource Registry, Massachusetts Eye and Ear Infirmary, 243 Charles Street, Boston, MA 02114
Toll-free phone: (800) 822-1327, Fax: (617) 573-3838, Email: tbregistry@meei.harvard.edu