



# the REGISTRY

*Newsletter of the NIDCD National Temporal Bone, Hearing and Balance Pathology Resource Registry*

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## GANGLION CELL LOSS IS NOT RELATED TO HAIR CELL LOSS IN HUMANS

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### Introduction:

To paraphrase a speaker at a recent ARO meeting, "Humans are not big mice". This is especially true when equating spiral ganglion cell loss to hair cell loss. There are numerous reports in the literature demonstrating that loss of hair cells leads to loss of ganglion cells in animals, but no investigations showing a similar effect in humans. In spite of the report by Nadol in 1989 that etiology of the hearing loss is the factor most closely related to ganglion cell loss [1], many papers, especially in the audiology literature in reference to cochlear implants, continue to state that hair cell loss leads to ganglion cell loss. These assertions are used to try and explain the poor performance of patients who are implanted many years after losing their hearing and to suggest that the recently introduced short electrodes, used in partial hearing loss, might cause further damage to hair cells and thus ganglion cells.

In this communication, we describe two studies that demonstrate that the loss of hair and supporting cells does not lead to a significant loss of ganglion cells in humans.



*See Ganglion Cell Loss is Not Related to Hair Cell Loss in Humans, page 2*

## Methods:

In the first study, we performed a morphological analysis of archival temporal bone sections and a statistical analysis of ganglion cell, hair cell and supporting cell populations.[2]

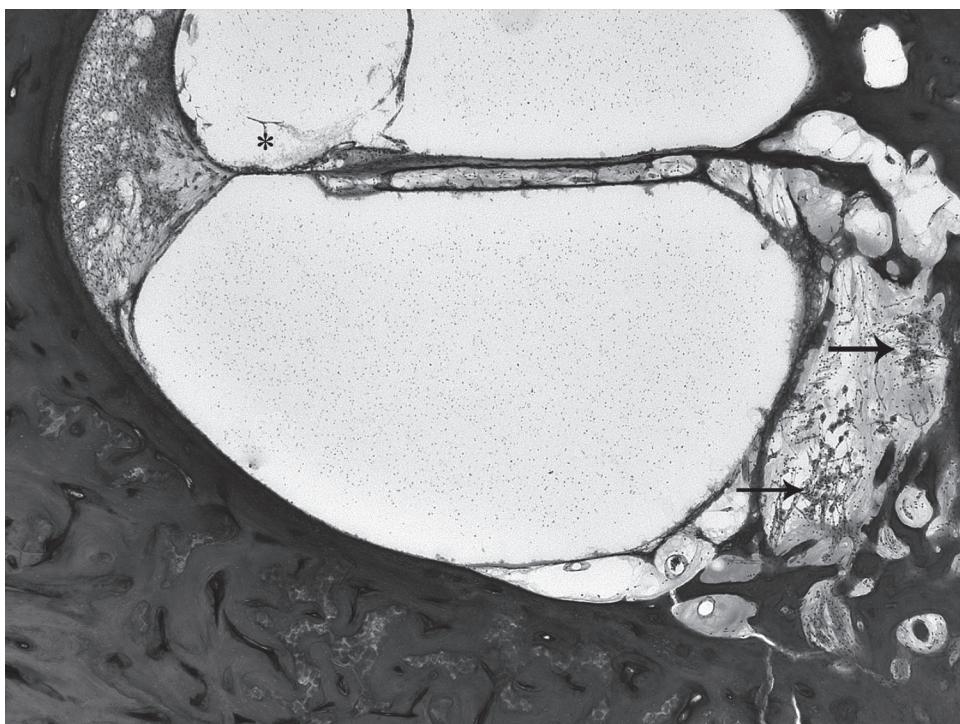
The second study comprised an analysis of four temporal bones that had a segmental loss of the organ of Corti limited to the cochlear base. Because of the small number of cases, cochlear reconstructions and cytococholeograms were used to investigate the relationship between hair cell loss and the ganglion cell population.



## Results:

Statistical analysis of archival specimens: In our collection of 1,442 temporal bones, there are 33 from individuals with total hearing losses of varying etiologies (labyrinthitis, otosclerosis, Ménière's disease, heredity, skull fracture, mastoiditis, cholesteatoma, and sudden sensorineural hearing loss) and duration (1 to 89 years). None of the ears had any residual hair cells. (Fig 1) Six ears were from patients who had received cochlear implants. The number of ganglion cells ranged from 34,299 to 2,889; the corresponding percentage of ganglion cells remaining based on age-normative data [3] were not significantly related to the duration of hearing loss ( $r=-.13$  and  $.02$  respectively,  $p>.05$ ) or to remaining supporting cell populations ( $r$ 's from  $.15$  to  $.27$ ,  $p>.05$ ). 51% of ears had ganglion cell counts within two standard deviations of age-normative means. There was no significant relationship of the presence or absence of peripheral processes (dendrites) and the number of remaining ganglion cells.

*See Ganglion Cell Loss is Not Related to Hair Cell Loss in Humans, page 4*



**Fig 1.** Photomicrograph from cochlea of a woman, deaf since birth, who died at age 89. Histopathological examination revealed a Sheibei deformity (cochleo-saccular degeneration). In spite of a loss of the organ of Corti (\*), the number of ganglion cells (arrows) was normal for her age.

# the REGISTRY

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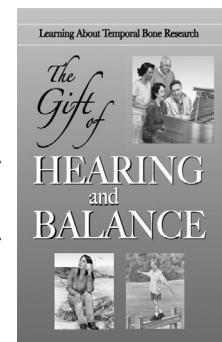
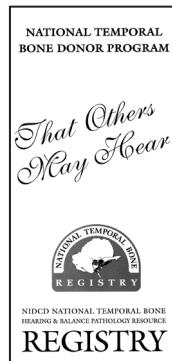
## Brochures about Temporal Bone Research and Donation Order Free-of-Charge for Your Office, Clinic or Organization

The NIDCD National Temporal Bone, Hearing and Balance Pathology Resource Registry, which is dedicated to promoting research on hearing and balance disorders through the study of temporal bones, has published two informational brochures, which you may request for display in your office and/or waiting rooms. Both brochures encourage individuals with hearing or balance disorders to bequeath their temporal bones to scientific research.

***That Others May Hear*** is a short form brochure which describes briefly the functions of the Registry, and answers commonly asked questions regarding the temporal bone donation process. (Dimensions: 9" x 4")

***The Gift of Hearing and Balance: Learning about Temporal Bone Donation*** is a 16-page, full-color booklet which describes in more detail and with diagrams, the structure of the ear, types of auditory disorders, the microscopic study of the temporal bone, and the benefits of temporal bone research. It also answers commonly asked questions regarding the temporal bone donation process. (Dimensions: 7" x 10")

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**.



## ORDER FORM

Please send me (circle or fill in quantity):

That Others May Hear:	25	50	100	<input type="text"/> copies (free of charge)
The Gift of Hearing:	25	50	100	<input type="text"/> copies (free of charge)
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Newsletters:	25	50	100	<input type="text"/> copies (free of charge)

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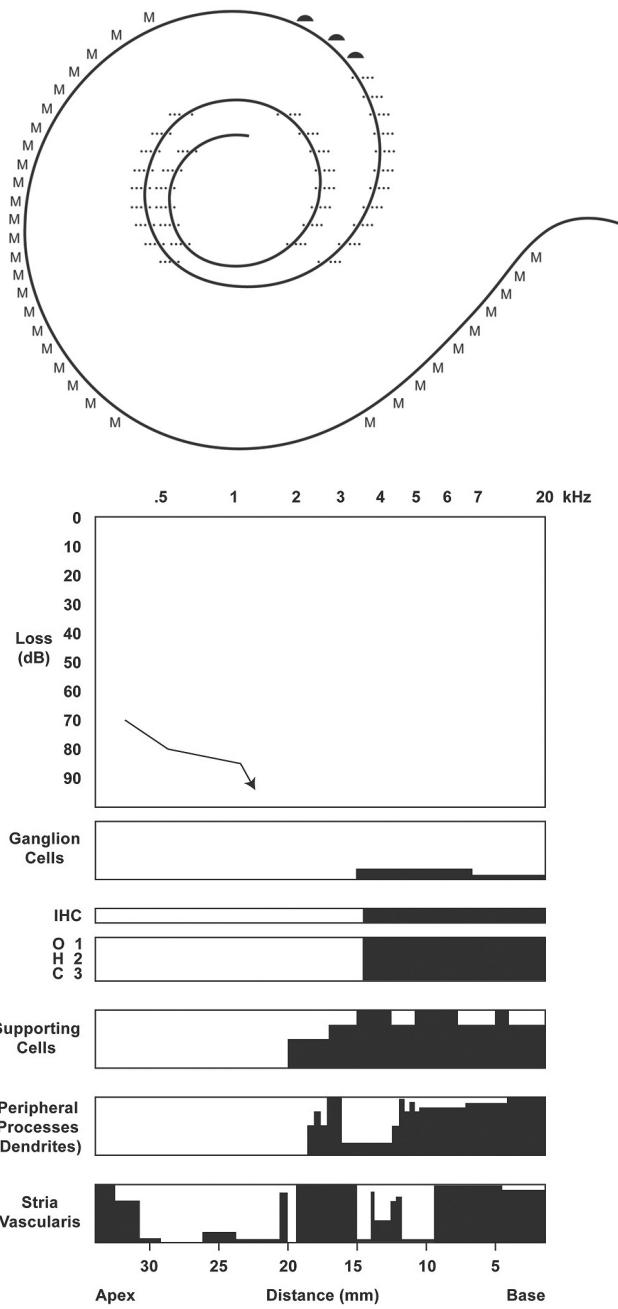
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Massachusetts Eye and Ear Infirmary  
243 Charles Street, Boston, MA 02114

(800) 822-1327 Toll-free phone, (617) 573-3838 Fax

Or submit your request using email: [tbregistry@meei.harvard.edu](mailto:tbregistry@meei.harvard.edu)





**Fig. 2.** Cochlear reconstruction of hair cells (top panel) and cytocochleograms (bottom panel) of 84 year old woman who suffered a sudden sensorineural hearing loss 15 years before death. In the top panel, M = missing Organ of Corti; black semicircles = Organ of Corti represented by mound of cells; dots = presence of hair cells. Although there was a total loss of hair cells in the basal turn, the loss of ganglion cells was minimal. The black areas in the cytocochleograms represent missing or abnormal elements. The inner and outer hair cells (IHC and OHC respectively) and supporting cells are recorded as present (white) or absent (black). Vertical axes for ganglion cells, peripheral processes and stria vascularis represent percentage of loss.

**Cytocochlear reconstructions:** Four additional cases were studied that had no surviving organs of Corti in the basal 15 mm extending from the round window toward the apex. However, the mean ganglion cell loss was only 25% compared to age matched controls. The ganglion cell loss in the apical region of these cases was 10%. (Fig. 2)

## Discussion:

Why there is a difference in the survival rate of ganglion cells after the loss of hair cells between humans and experimental animals is, as yet, not known. It might be hypothesized that the life span of most animals is shorter than humans and, therefore, the changes occur more rapidly in animals. However, a review of the literature indicates that the percent of the life span between the loss of hair cells and ganglion cells is 1% for chinchillas, 8% for cats, 25% for guinea pigs, but indefinite for humans.

There are two anatomical differences between human and animal ganglion cells. Most human ganglion cells are unmyelinated, in contrast to animals; and there are synaptic and junctional interactions between the cells in humans.[4, 5]

The determination that loss of hair cells will not lead to loss of spiral ganglion cells should put to rest the concept that the poor performance of deaf patients implanted late in life is due to ganglion cell loss resulting from long-standing hair cell loss. In fact, there is evidence that the ganglion cell population has little to do with performance. [6, 7]

Gantz has introduced the use of short electrodes to supplement high-tone hearing loss with a cochlear implant to stimulate the basal areas of the cochlea.[8] There has been concern that this might damage surviving elements of the organ of Corti and lead to further loss of neural elements [9,10]. The finding that loss of hair and supporting cells does not lead to loss of ganglion cells should allay these fears.

## References:

1. Nadol JB, Young YS, Glynn RJ. *Survival of spiral ganglion cells in profound sensorineural hearing loss: implications for cochlear implantation*. An Otol Rhinol Laryngol, 1989. **98**: p. 411-416.
2. Teufert KB, Linthicum FH Jr, Connell SS. *The effect of organ of Corti loss on ganglion cell survival in humans*. Otol Neurotol., 2006. **27**(8): p. 1146-51.
3. Schuknecht HS. *Pathology of the Ear*. Second ed. 1993, Philadelphia: Lea & Febiger. p. 25.
4. Nadol JB Jr. *Comparative anatomy of the cochlea and auditory nerve in mammals*. Hear Res, 1988. **34**(3): p. 253-66.
5. Tylstedt S, Kinnefors A, Rask-Andersen H. *Neural interaction in the human spiral ganglion: a TEM study*. Acta Otolaryngol, 1997. **117**(4): p. 505-12.
6. Nadol JB Jr., Eddington DK. *Histopathology of the inner ear relevant to cochlear implantation*. Adv Otorhinolaryngol, 2006. **64**: p. 31-49.
7. Fayad JN, Linthicum FH Jr. *Multichannel cochlear implants: relation of histopathology to performance*. Laryngoscope, 2006. **116**(8): p. 1310.
8. Gantz BJ, Turner CW. *Combining acoustic and electrical hearing*. Laryngoscope, 2003. **113**(10): p. 1726-30.
9. Kiefer J, Gstoettner W, Baumgartner W, Pok SM, Tillein J, Ye Q, von Ilberg C. *Conservation of low-frequency hearing in cochlear implantation*. Acta Otolaryngol. 2004; **124**(3): p. 272-80.
10. Scarpidis U, Madnani D, Shoemaker C, Fletcher CH, Kojima K, Eshraghi AA, Staecker H, Lefebvre P, Malgrange B, Balkany TJ, Van De Water TR. *Arrest of apoptosis in auditory neurons: implications for sensorineural preservation in cochlear implantation*. Otol Neurotol. 2003; **24**(3): p. 409-17.



## 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
- 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.  
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243 Charles Street  
Boston, MA 02114



## NIDCD National Temporal Bone, Hearing & Balance Pathology

### Resource Registry

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## News and Announcements

***Look for the Registry's Exhibit at  
these upcoming meetings***



The Registry will be exhibiting at the upcoming AARP Life @ 50+ National Event and Expo in Boston, Massachusetts from September 6th-8th, 2007.

[www.aarp.org](http://www.aarp.org)



The American Academy of Otolaryngology -- Head and Neck Surgery 2007 Annual Meeting & OTO EXPO will be held in Washington, D.C. on September 16th-19th, 2007.

[www.entnet.org](http://www.entnet.org)

### Did you know?



The NIDCD Information Clearinghouse has many publications available to patients and physicians on various topics related to hearing and balance. The list of publications is available on their website at [www.nidcd.nih.gov](http://www.nidcd.nih.gov).