

Hearing Health

Winter 2014

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SPECIAL MUSIC ISSUE

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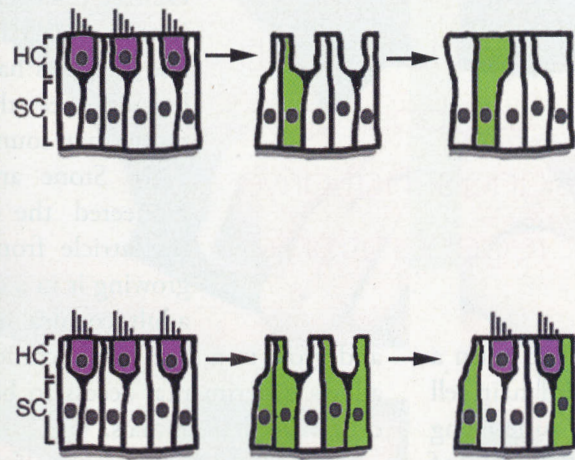
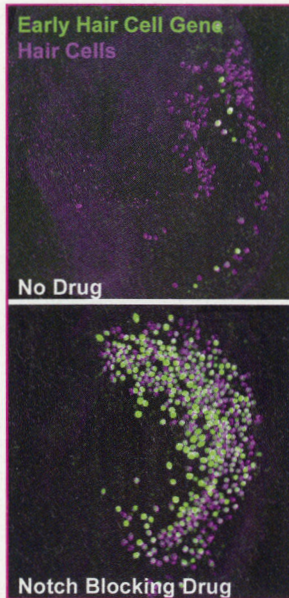


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The lab of Jennifer Stone, Ph.D., has developed a way of growing the adult mouse utricle in culture. The top far left panel shows an adult utricle in which most of the hair cells (depicted in purple) have been killed with drugs. A few supporting cells (SC) have begun to turn on early hair cells genes (green), but they cannot fully convert into hair cells (HC). This is schematized in the illustration on the immediate left.

The bottom far left panel shows a similar mouse utricle, but this time grown with a drug that blocks Notch signaling. Many more supporting cells have turned on early hair cell genes (green), and many of these cells have gone on to convert into hair cells.

In this scheme, supporting cells make a protein on their cell surfaces. This is the Notch receptor. Hair cells make proteins on their cell surfaces that bind to the Notch receptor, like a key fitting into a lock. Binding to the Notch receptor is believed to actively block supporting cells from turning into hair cells.

Dr. Stone and her team found that if they killed hair cells in the utricle, but then used drugs to block Notch signaling, they saw small but significant numbers of true hair cells appearing in their experiments. These results suggest that the Notch pathway may be one of the signals that holds regeneration back in the adult mammal.

Neil Segil, Ph.D., and I have teamed up with Dr. Stone and her lab to identify other players that may block regeneration in the utricle. Using Dr. Stone's culture

system, our three labs will use next-generation DNA sequencing technology to identify genes that are switched on in supporting cells of the utricle as they attempt to transform themselves into hair cells after damage—but ultimately fail. We will then see what genes are switched on or off when drugs that block the Notch pathway are used to finally complete the process and drive supporting cells to produce hair cells.

These experiments are still at a very early stage, but we hope this approach will complement other work by members of the HRP consortium that are looking at similar processes in the cochlea.

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A Grand Passion



A pianist and Hearing Health Foundation board member explains why the Hearing Restoration Project is important to her—and to everyone with hearing loss.

By Nancy M. Williams

For years, I tried to consider my hearing loss as little as possible. I never dreamed there would be a cure for hearing loss and tinnitus. Now as a board member of Hearing Health Foundation (HHF), I am part of a team working toward a cure. This new role has forced me to confront the societal stigma around hearing loss and, even more deeply, my own denial.

In kindergarten, after I sang "Three Blind Mice" too loudly on the big rag rug in our classroom, I was diagnosed with a high-frequency hearing loss. My parents, worried about the social stigma, refused the recommended hearing aid, a decision that boomeranged when I reached middle school. "You can't hear secrets," complained a girl with green eyeliner at lunchtime.

"Don't sit with us anymore."

I was devastated. My parents broke down and had me fitted with an aid, a behind-the-ear model, bulky by today's standards.

My parents had acted with the best of intentions in a society that tolerated hearing loss even less than ours does today. Yet the incident in the lunchroom stayed with me for a long time. To compensate, I rarely admitted to anyone that I had a hearing loss. By some tacit pact, my closest friends, who knew about my loss, rarely mentioned it. The shame I felt in the lunchroom had mildewed within.

Once I was fitted with that first aid, however, I never stopped wearing it. Over the years I upgraded to two aids, then two digital aids, until I arrived at a high-performance, six-

setting pair of aids that included a special music setting. As a passionate amateur pianist, I felt it was only natural that two of the settings be for music—one for practice, the other for performance. Although my hearing aids were on the cutting edge of technology, I still consigned thoughts about my loss to a remote, secluded part of my mind.

Then, in the fall of 2011, a friend asked me to cover a research symposium hosted by HHF. I attended as a member of the press, but as I rode the elevator up to the symposium, I felt no wiser than my little self on that big rag rug in kindergarten. I felt tumult at the prospect of walking into a roomful of people with hearing loss. I pictured people with ear trumpets,

as cranky as deaf Beethoven, yelling, "WHAT?" The truth was, I had bought into society's depiction of people with hearing loss; I was reluctant to confront the part of me I usually tried to deny.

Five minutes into the reception, my fear morphed into elation that didn't dissipate the entire night. I met professionals with hearing loss, parents of young children with hearing loss, well-regarded doctors who treat hearing loss, and most importantly, some of the scientists who are part of the Hearing Restoration Project (HRP). The mildew within—the shame I felt for my hearing loss—began to dry up and disappear.

In volunteering with the foundation, I learned more about the HRP consortium of scientists, who, contrary to the tradition of competing against one another and racing to be the first to publish their findings, had agreed to work together. I admired the scientists' approach and joined the foundation's board.

Now I am in the hands of the HRP. Since my loss was first diagnosed, the curve of my hearing has been

slipping downward on the audiological charts. Now, in the conversational frequencies, my hearing is no longer normal—in fact it has slipped past a mild loss to moderate, and is sloping down to severe in the high frequencies. Sometimes I worry about the sounds that someday I might miss: the treasured voices of my husband and children and the sweet chimes and deep rumbles of my piano.

I need the HRP to succeed. We need the HRP to succeed—people with hearing loss and tinnitus of all ages and backgrounds and beliefs and professions need the HRP to succeed. This is true especially of children, some of whom—despite all the progress our society has made, signified by colorful "look at me" aids and cochlear bling—may still be shunned in the lunchroom.

Although it is off to a very promising start (see Dr. Groves's accompanying story), the HRP is still in its early days. The scientists need additional funding in order to create



momentum with their research and establish a cycle that will lead to success. Abolishing the stigma around hearing loss goes hand in hand with securing funding.

I hope you'll join me in advancing both of these causes, by talking candidly and passionately about your or your loved one's hearing loss, and by donating whatever you can, no matter how small, to the HRP. I had never dreamed there might be a cure for hearing loss, but now I have hope.

Nancy M. Williams, who serves on HHF's board of directors, is the founding editor of *Grand Piano Passion*, an online magazine for musicians with hearing loss. To learn more, see grandpianopassion.com.