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Caught Up in a Whirlwind of Genetic Hearing Loss

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Caught Up in a Whirlwind of Genetic Hearing Loss



BY BRENDA BATTAT

I was born into a family with genetic hearing loss going back at least four generations. That's as far as I have been able to trace it even though I have the family tree dating back to the 17th century.

Fifty-six years ago, when I was first diagnosed with moderate hearing loss at the age of 19, I was told it was a dominant genetic condition that could be passed down by one parent alone and that each child had a 50 percent chance of inheriting the gene. It was a roll of the dice with each pregnancy.

It certainly gave me pause when deciding whether or not to have children. But I rationalized that I was glad I had been born. It's certainly a challenge living with hearing loss, but then again don't we all face challenges?

I also know that things are so much better now than they were 70 years ago for my grandfather when he was living with hearing loss. He found solitude in his English garden and walks in the cemetery where no listening demands were made on him.

I wrote about the difference 70 years makes between my grandfather's and my own experience in the July/August 2006 issue of *Hearing Loss Magazine* after I received a cochlear implant. Today we are light-years ahead; so much more information is available, accessibility through technology is evolving at an unprecedented pace, and there are discrimination laws providing rights and protections unheard of in my grandfather's time. Current and future generations have more tools at their disposal to live and work successfully with hearing loss than ever before.

The Family Tree Keeps Growing...

I have two children—a boy and a girl. My son, James, fell into the family pattern, being diagnosed with hearing loss in his late teens. Upon seeing his audiogram my rationalization for having children was overtaken by emotion, and I can't pretend it didn't shake me up mightily. I was shattered; it was the reality of a worst fear. He was the one consoling me, "Mom, it will be all right." By this time James had attended several HLAA Conventions with me and had been around many HLAA Members, and I am sure all those role models lessened the blow for him.

Years passed. James started to use hearing aids when he was in his twenties. We assumed my daughter, his older sister Anna, had escaped the gene. However, when she was in her early forties, she noticed that she was having trouble hearing one of her children in the back of their car, so she decided to get her hearing checked.

Anna's audiogram not only showed she had a hearing loss but one at the same level as her brother, who had been using hearing aids for years. She hadn't noticed any difficulties communicating nor had any of her friends, co-workers or family. How hard her brain must have been working all these years to make sense of what she was hearing!

...And Growing

So Anna had also inherited the gene after all. She immediately decided to have her two boys—8-year-old Teddy and 9-year-old Leone—tested. Even though they had both passed the newborn hearing screening it turned out Leone now had a hearing loss.

Anna's experience pushed James to have his three children—Julian (3), Kiernan (7), and Sebastian (9)—screened, as they had also passed the newborn hearing screening. His expectation was that if they inherited the gene from him it would show up in their late teens, which is why he had delayed testing them. But no, now Sebastian was diagnosed with hearing loss. As I write this we are awaiting the results for Julian.

Within a two-month period, three members of our family had been newly diagnosed with hearing loss: Two of my five grandchildren and my daughter. It felt like we were all caught up in a whirlwind.

Sadness Turns to Resolve

Passing on the gene to my two children was bad enough, but I cannot describe the feelings when my adorable grandchildren were diagnosed with the family's genetic sensorineural hearing loss. I felt I was bringing intense sadness into our family. I had never really thought far enough ahead to my grandchildren, even though the possibility that they would inherit hearing loss was very real. But so early—nine years old? The pattern in

Brenda's daughter, Anna, with her husband Benedikt and sons Teddy (left) and Leone.



James Battat, holding son Julian, with wife Erin, son Sebastian and daughter Kiernan.



Left photo—Brenda’s grandchildren Leone and Teddy; center photo—Brenda with her husband Joe; right photo (l to r)—grandchildren Sebastian, Kiernan and Julian.

our family had always been late teens or early twenties, but now I realize that was when we were diagnosed, not when the hearing loss actually started. We are now picking it up earlier due to a heightened awareness.

Once I pulled myself together, I decided this has to stop. When I was told we had a dominant genetic condition it was all the information available at that time. Since then the field of genetics has evolved dramatically, with more than a hundred mutations for hearing loss having been discovered. There is also exciting work being done in the area of genome editing—removing the mutation and disabling the gene.

My deep, deep sadness turned to resolve to rid our family of this cycle; to stamp it out. But first we had to see if we could identify the specific gene causing our problem. I researched the most promising and exciting genetic research programs available and enrolled all of us into one.

My immediate and extended family members—many of whom live in England—were officially accepted to be part of a research program at the Molecular Otolaryngology and Renal Research Laboratories (MORL) at the University of Iowa. The MORL has developed the most comprehensive platform that screens all known deafness-causing genes (152 genes and microRNAs) and provides an accurate molecular diagnosis for people with hearing loss. This is combined with a study of the family history and audiograms that we collected on both sides of the Atlantic. I am hopeful that we will learn the cause of our family hearing loss and be in a better position to benefit from advances in gene editing, perhaps even finding a cure in my grandchildren’s lifetime.

Our New Normal

We are all gradually adjusting to this new reality—grandparents, parents, and children. We now have a total

of seven hearing aids and one cochlear implant among us! It is really impressive how maturely the young children are dealing with their hearing loss both at home and at school. When Sebastian saw the dip in his audiogram he asked the audiologist some very hard questions, “Does it go down further? Will it go back up?” Out of the mouths of babes. He is not a candidate for hearing aids yet.

His cousin Leone is already wearing two navy blue hearing aids and has a remote microphone that connects via Bluetooth. He handles them like a pro—changing the batteries, putting them in a dry pack at night, and changing the channels to suit his environment.

Leone is fortunate to be supported by an impressive school system and school audiologist. They have been proactive and very accommodating, including doing what they can to minimize background noise in the classroom—putting tennis balls on the feet of chairs, lowering interference from the HVAC, improving room acoustics, teachers using a microphone, adjusting seating arrangements, allowing him to take fatigue breaks from listening during class, captioning of videos, and providing the context of class discussions to him up front so he can anticipate the vocabulary that is likely to be used. They are open to other accommodations as identified and needed by Leone.

What a difference inclusion in the classroom has made. Students are used to different learning styles and needs and don’t find it odd that their classmates may need certain accommodations. In fact, they are most likely benefiting from many of the accommodations themselves. Such things as captioning, reduced classroom noise, and sound field amplification have all been shown to facilitate learning for all. And for Leone it means that he is willing to accept using hearing aids, microphones

and generally doing whatever it takes to make it easier for him to fully participate in the classroom.

How Far We Have Come!

Contrast the classroom experience of my grandchildren with that of my own. Even though I had not been diagnosed with hearing loss at the time I was their age I always selected a seat at the front—not typical of kids who would most likely prefer to be as close to the back as possible. Our hearing was tested at school by listening to the ticking of a wristwatch. I never actually heard it, although I said I did. My internal rationale was that if they had held it there longer I would have been able to hear it.

It wasn't until I was 19 years old when I couldn't hear whispered instructions in a very busy orthopedic clinic did my supervisor and I both suspect I had a hearing problem. That was when I went for my first real hearing test. I remember it clearly; I was told I had a sensorineural hearing loss, and that given my family history it was probably progressive. I was also told that since the loss was in the higher frequencies I would “fortunately” be able to hear men's voices better than women's. Imagine being told that in today's environment!

As we await the results from the genetic screening we are undergoing in Iowa I am still sad, but also expectant and hopeful because of the heightened attention to hearing loss as a public health concern. For far too long the impact of hearing loss has been underestimated by policy makers, researchers, medical professionals and individual consumers themselves who fail to seek treatment even when they suspect they have hearing loss.



THE MOLECULAR OTOLARYNGOLOGY AND RENAL RESEARCH LABORATORIES

For more information about the Molecular Otolaryngology and Renal Research Laboratories (MORL) at the University of Iowa, visit them online at:

-  morl.lab.uiowa.edu
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Two key reports that have come out recently—one from the President's Council of Advisors on Science and Technology (PCAST) and the other from the National Academies of Science, Engineering, and Medicine (NAS)—have shaken up the status quo by offering recommendations to expand technology options for consumers, improve services and boost treatment outcomes for people with hearing loss.

In August 2017, the Over-the-Counter (OTC) Hearing Aid Act was passed, which will enable adults with mild to moderate hearing loss to purchase FDA-regulated hearing devices without being seen by a hearing professional. We are awaiting for the FDA (Food and Drug Administration) to release regulations to ensure high standards of safety, labeling and manufacturing protection. More recently the Centers for Disease Control (CDC) discussed causes of hearing loss and prevention strategies and public health solutions as part of its Public Health Grand Rounds.

This heightened attention and awareness is stimulating research to build on what we already know—that untreated hearing loss can lead to isolation and depression, up to a threefold increase in the risk of falling in older adults, more hospital stays, and possibly even dementia.

As the search for more and better treatments—even cures—continues, I believe genetics is going to play a big part in the solution, maybe not in my lifetime but I'm willing to bet in my grandchildren's. **HL**



Over the past 30 years Brenda Battat has worked in the field of hearing loss—as a staffer and executive director of the Hearing Loss Association of America (HLAA) until 2013, and as an advocate and consultant in an array of government, nonprofit, industry and advocacy settings. Brenda has served on many advisory boards related to hearing and hearing science. In recognition of her stature as an advocate for people with hearing loss, Brenda served as the key representative for consumers on the National Academies of Sciences, Engineering, and Medicine (NAS) Consensus Committee on Affordable and Accessible Health Care and the advisory council of the National Institutes of Health (NIH) National Institute on Deafness and Other Communication Disorders (NIDCD). She has also served as an expert witness in court cases related to hearing issues. Brenda received her undergraduate degree in physical therapy from St Mary's Hospital in London and her master's degree in counseling from Indiana University. She has lived and worked in four countries and taught English in China for two years.