Question response: Webinar: 8 November 2021

Professor David Ryugo – Gavin Institute

Hearing Loss and the Brain: Making Sense of Sound and Hearing Loss

- **Stem cells - they are used for generating new cells, are they potentially able to be used to repair damaged ears (like they use them for sports injuries for example).**

  Stem cells offer the potential to "replace" injured or missing cells and to repair tissue. To date, this potential is still in the experimental stage. One can read about replacement of cartilage, ligaments, muscles and tendons but these tend to be in advertisements and in foreign countries, not in Australia. The treatments also do not last very long, and claims of fraud are common. Basically, stem cell therapy is still in its infancy.

  Research has shown that when stem cells are applied to the inner ear, there are new sensory receptors generated but they are distributed all over the inner ear, not in the specific location where they belong. Recall the orderly array of sensory cells--3 rows of outer hair cells and 1 row of inner hair cells--that is precision architecture. The ear is heaps more complicated in structure compared to a tendon, ligament, or cartilage. When scientists are able to direct the stem cells to the proper location, then we might have a remedy for hearing loss using stem cells. At present, however, it is not practical. Things to think about: (1) use cells that regenerate and multiply creates a situation that resembles cancer growth; (2) stem cell reproduction has to be controlled; (3) stem cells have to go only where they belong; (4) once in the ear, stem cells have to become connected to the auditory nerve fibers that take the information into the brain, so controlling how fibers and sensory cells connect is another HUGE challenge; (5) the sensory cells reside on the flexible basilar membrane, which is mechanically "tuned" to become excited in different places depending on the frequency of the incoming sound vibrations--this tuning is exquisitely balanced. Any additional mass brought by extra stem cells will upset the "tuning" and probably cause a different part of the membrane to vibrate. The sound vibrations would therefore activate the wrong part of the ear so neural excitement would not be properly linked to the sound--you could lose frequency tuning of the ear.

- **Why do people with Meniere's get pain/ pressure in their ears**

  We don't know what causes Meniere's disease but we know that the presence of MD results in an overabundance of fluid in the fluid-filled compartments of the inner ear. The inner ear is bone that is shaped into (1) the snail-shaped cochlea for hearing; (2) a complex of 3 semi-circular canals oriented in the lateral, vertical, and horizontal planes; (3) and a vestibule that links the cochlea and the canals. This structure is called the bony labyrinth. Inside this bony structure is the membranous labyrinth, a compartment with membrane walls within which reside the sensory cells for hearing and balance. The different clusters of sensory cells are well separated but lie within the same compartment of fluid. Meniere's Disease is where there is too much fluid in the membranous labyrinth--either because of over-production of fluid or insufficient drainage of the fluid, causing the membrane to swell. It is thought that the abundance

of fluid is what causes the "sense of fullness" as well as hearing loss and dizziness. It is not known why the fluid volume fluctuates or what causes it. Medication that increases water excretion (diuretics) is often used to relieve the inner ear of fluid build-up. Low salt diets are also encouraged.

- **Why do we lose different hearing frequencies, rather just have blanket hearing loss. You said older people lose high frequency, while Meniere's people lose low frequency?**

  Hearing is a complicated business. And one can have partial damage--resulting in limited hearing loss, or major damage--causing deafness. So catastrophic damage--a genetic mutation affecting the ear, extremely high fever such as with mumps or measles, or certain drugs--can produce a range of hearing loss: from a little to total deafness. Remember, the sensory receptors sit on a membrane that vibrates in different locations, depending on the frequency of the sound. So different sounds activate different parts of the inner ear. Really loud sounds will damage that part of the membrane plus the sensory cells where excess vibration occurs. This restricted damage will cause restricted hearing loss. Noise is the most common source of restricted damage--turning the sound of your headphones up too loud, working around power tools without ear protection, driving a really loud motorcycle, using a loud hair dryer or vacuum cleaner or coffee bean grinder--these sources of noise trauma will give you regions of damage causing restricted loss of hearing at the frequencies where the damage occurs. For reasons we don't fully understand, the high frequency part of the inner ear is especially vulnerable to all loud sounds.

  An analogy--nearly everyone born with normal vision will nonetheless begin to need reading glass at some point after they turn 40 years of age. The reason is because as we age, the once very flexible lens in the eye becomes stiffer. It means that our visual system can no longer accommodate viewing close-up objects without help. Reading classes provides that help.

  In the ear, we have the sensory receptor cells residing on a differentially flexible membrane, called the basilar membrane. Normally, the part of the membrane nearest the vestibule is narrow and relatively stiff--perfect for vibrating at high rates as when high frequencies are delivered to the ear. As we move away from the base of the cochlea, the basilar membrane becomes "looser" so it can vibrate at lower frequencies. At the tip of the cochlea, furthest from the vestibule, the basilar membrane is maximally loose and floppy. It is thought that when the inner ear membrane starts to overfill with fluid from Meniere's disease, the effect of the increased pressure occurs where the structure is "loosest"--that is, where low frequencies are processed. This pliability of the membrane at the apex may be why low frequencies are preferentially affected.

Question response: Webinar: 8 November 2021

- **What is the impact of hearing loss to other functions that the brain carries out when it is compensating and using extra energy to process damaged hearing signals?**

  When someone struggles to hear and/or understand--when you have hearing loss or when you don't understand the foreign language being spoken to you--you must exert tremendous mental energy to concentrate in an attempt to understand. It is this intense concentration and (possibly) self-conscious worry about missing something important or appearing stupid that makes us mentally fatigued.

- **What is the significance of the inner and outer Hair cells? Is there an afferent and efferent pathway to the Cochlea?**
  We have 2 sets of sensory hair cells, also called receptor cells. There are 3 rows of outer hair cells and 1 row of inner hair cells. The outer hair cells are thought to be involved in actively vibrating "in harmony" to the incoming vibrations on the basilar membrane to amplify the incoming sound. The inner hair cells vibrate to the incoming sound and convey the frequency information to the auditory nerve fibers, which carry sound information into the brain. Fibers that connect to hair cells and project into the brain (afferent fibers) convey the sound information to brain cells. These afferent fibers initiate the auditory pathways in the brain that ascend to the cerebral cortex to yield perception (meaning) of the sounds.

  Remarkably, the brain has a separate circuit that originates in various auditory structures and sends neural information downwards--this feedback loop seeks to control the information coming up from the cochlea, and is thought to be regulated by the cerebral cortex. The cortex tells the lower centers to turn the gain up or down, depending on the sound and the situation. It can also work to make sounds "sharper" by inhibiting hair cells at the edge of the stimulated zone.

  At the bottom of this descending pathway reside the cell bodies of the efferent fibers. These fibers project from the brain to the sensory receptors of the inner ear. Projections to the outer hair cells turn the sensitivity of the ear up or down; projections to the inner hair cells modulate the sensitivity of the auditory nerve fibers. Together, the efferents work to turn down the gain when in an environment of loud noise or to help with selective listening.

- **A problem for us is the direction of sound? We can't work out where it is coming from. Why is this?**

  Sound localization requires the brain to perform some heavy-duty computations. For touch or vision, the sensory epithelium--the receptor sheet--is simple. For touch, the sensory surface is the skin. The skin is represented by the various brain structures processing touch--and it knows "space" or the area of stimulation by virtue of the brain map of the skin. Likewise, the visual receptor surface in the eye, acts like a photographic plate and the connections within the brain replicate the visual space. The cues for sound space are brought into the two ears--they determine the time and loudness differences between the two ears to calculate where in space the sound originate. Trouble distinguishing the origin of a sound in space could be due to an imbalance

between the two ears. If that is the problem, then balancing the ears using hearing aids could help, or simply practicing listening in a quiet room could help. You might have to relearn the signals--that would recalibrate your internal space map--to improve on sound direction. Or it could be something more serious, and you should visit a neurologist.

- **So noise cancelling earphones are probably REALLY important for us to help save our hearing?**

  Noise-cancelling earphones are good for somethings, not everything. They are good when the environment is loud but monotonous; the built-in software can cancel noise. A good example is when you are flying--the background noise gets cancelled. A different kind of noise protection is something called a low-pass filter. Such a headphone is used in the military to block sudden onset sounds, such as gunfire. Sounds that are not so abrupt are allowed through. Noise-cancelling earphones do not work where the background is organized, such as speech. The time varying speech means that the system doesn't know what the noise is. The best response to severe noise is to block out all loud sounds.

- **Over the past 6 months I have been experiencing daily fluctuations in my hearing and tinnitus. One day my hearing is normal by the afternoon I have moderate loss and wake up with severe loss over a few days then back to normal and the cycle starts again. What does this type long term hearing fluctuation have on the cochlear and the brain?**

  Fluctuating hearing loss and/or dizziness is standard for Meniere's disease. In a general way, it is attributed to fluctuating levels of fluid within the membranous labyrinth. The cause of these fluctuations, while unknown, has been speculated to be due to viral infection, autoimmune disease, or inflammation. Maybe stress or even something in the diet. You could keep a daily diary of what you eat, how stressed you feel, your general activities, etc. to see if you can't find some relationship to what you are doing that makes your tinnitus/hearing loss act up.

- **What about hearing issues like transference, which I have? Even when my hearing aid assists with hearing sound, I can’t understand what is being said properly.**

  I'm sorry but I don't know about the term "transference." But having "OK" hearing thresholds but difficulty understanding speech is the classic example of what is becoming known as "hidden hearing loss" in adults or "auditory neuropathy" in children. You would be advised to see an ENT doctor or a neurologist who specializes in hearing cognition.

Question response: Webinar: 8 November 2021

- **Can a Cochlear implant reverse changes in the brain?**

  It is certainly clear that a cochlear implant can reverse or prevent many brain pathologies resulting from hearing loss. You just have to witness how well an early-implanted child functions after a few years of using the device. Brain changes caused by untreated hearing loss affects young people far greater than when it occurs in adults (after puberty). It is best to be pro-active when thinking about hearing loss and brain changes: always attempt to remedy your hearing loss (hearing aids or cochlear implant) to maintain socialization skills and to prevent or postpone any accompanying brain change. Prevention is best, when possible.

- **Does Meniere's destroy the receptors? Do we know how this happens?**

  Meniere's disease can destroy hearing and balance receptors if the fluid volume increases within the inner ear to the point of rupturing the membranes holding the fluid and the sensory receptors.

- **So much of my issue with Meniere is ear sensitivity. I notice when you say "hot" my ear is ok but when you say "hat", my ear tenses and hurts. So I suppose I'd say higher frequencies upset my ear (and make me feel dizzy) which is so different than hearing loss. Why might that be?**

  I've found the same thing, has taken a few goes to get it right. At first sounded tinny and loud, now it's so much better.

  These last 2 statements seem to address what we'd call "hyperacusis" a super-sensitivity to sudden changes in loudness. If you look at the spectrogram of these words (link below)

  https://auditoryneuroscience.com/vocalizations-speech/formants-harmonics

  Notice that "hat" has much more energy in the vowels compared to "hot"--that extra energy could be the source of your sensitivity. The high frequencies in the consonants (the right hand band of energy in each spectrogram mostly missing in the low pitched voice saying "hat") might be sufficiently low that you aren't bothered by them. The word "head" might also be annoying.