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Dr. Terry-Lynn Young, Memorial University

'A solution for hearing loss – if only people would listen"

At 12- or 13-years of age, a good student turns bad. He doesn't seem to listen in class. He's restless.

The boy in this story is a real one. He grew up in an isolated community on the South Coast of Newfoundland, where the genetic lines run deep through many generations.

Dr. Terry-Lynn Young, a member of the gene dream team at Memorial University in St. John's, first heard about the lad more than 20 years ago.

"He started acting out." A hearing test showed he had trouble hearing low frequency sounds. "He had a male teacher, and he simply couldn't hear him well."

It turned out the hearing loss was a genetic legacy – and the cause of the sudden onset of his troubles at school.

Young was a postdoctoral fellow and member of the Memorial team that worked with members of the boy's family, several of whom wore hearing aids.

Anne Griffin, an audiologist on Young's team confirmed low-frequency hearing loss in family members. Unfortunately, the hearing aids had been tuned to amplify high-pitched sounds.

The audiologist adjusted the aids, cranking up the volume of low frequency sounds. "Grown men came out of her office with tears in their eyes. They could finally hear," says Young, a leading geneticist who spent her formative years in St. Anthony, at the top of Newfoundland's Great Northern Peninsula.

The Memorial team conducted genetic testing, finally finding the gene that had impacted the family's hearing for eight generations.

In 2001, Young and others published the results in *Human Molecular Genetics*, a journal published by Oxford University Press. Almost two decades later, she is "incredibly frustrated" that the key insight of that peer-review article has not made its way widely into clinical practice.

The insight was that a patient's genetics should form the basis of care for hearing loss – from each according to his genome, to each according to his needs.

Care for hearing loss, based on genetic testing, is not complicated as it sounds, either. In some cases, audiologists can simply fine tune hearing devices the same way you do your stereo – turn down the bass, turn up the treble, and deliver more sound to the right speaker if required.

Suite 403
5657 Spring Garden Road
Halifax, Nova Scotia
B3J 3R4
902-425-4230
Fax: 902-425-4233
www.atlanticuniversities.ca

But the genetic understanding of hearing loss now goes deeper than that, Dr. Young said.

“We can tell if the hearing loss is in one ear or both ears, if it gets worse or stays the same over time, if it is high frequency or low frequency loss, if it is likely to impact people when they are older or younger. It’s like we know each and every cell.”

The underlying health care issue is urgent. Hearing loss is “the most common sensory disorder of all in humans”, she says, and is now linked to social isolation, dementia in older people, and learning problems in children.

Young describes the cochlea - the snail shaped portion of the inner ear that converts sound vibrations into nerve impulses to the brain – as “elegant” and beautiful”. It’s a triumph of genetics that the cochlear system is so well-understood. It’s a failure of health care delivery that this understanding isn’t better reflected in clinical practice.

A good start at improving care delivery would be easier public access to genetic testing, which can be done from a single strand of hair or saliva sample just like on a TV cop show. “How do we roll out genetic testing at Costco?,” Young asked.

Another step forward would be better training in genetics for student audiologists. That’s sorely lacking today. As a result, audiologists use standard hearing tests, which often fall short of what is needed and as the family from the South Coast of Newfoundland learned the hard way.

As part of Memorial’s team of geneticists, Young knows that genetic breakthroughs can sometimes result in better patient care in a hurry.

She and her colleagues identified a genetic anomaly in a Newfoundland family that caused sudden cardiac arrest and often early death – especially among males. “Your first symptom could be your last,” she said.

Cardiologists implanted defibrillators in these patients - to restart the heart in case of an incident – before Young’s team found the underlying gene.

That was “brilliant’, she said. The caregivers were out in front of the genetic knowledge, not trailing it.

Young is determined to see genetics play a more significant role in treating hearing care as well.

“I’m going to beat this drum loud enough that everyone hears” she said.

Research That Matters is written by Jim Meek, Public Affairs Atlantic on behalf of the Association of Atlantic Universities (AAU).

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