



## Researchers correcting recessive genetic deafness

Two years ago, David Liu repaired a dominant mutation and prevented hearing loss in a mouse model for the first time. A breakthrough - but, most genetic diseases are not caused by dominant mutations. They're caused by recessive ones, including most genetic hearing loss. Now, Liu and researchers have achieved another first: Successful genome editing to fix a recessive disease-causing mutation.



## Free aged care Auslan interpreting service

The Australian Government has launched a free national sign language interpreting service for senior Australians who communicate in Auslan and receive or want to access aged care services. The Minister for Aged Care said, "It will ensure that these people can participate in the assessment, planning, and review of their care."



## Marie was just four-years-old when she knew she wanted to be a teacher

So began her path to eventually open WA's first school for deaf children. Marie Kormendy was recently appointed a Member of the Order of Australia for her "significant service to people who are deaf or hard of hearing through humanitarian assistance programs".

## Acoustic weapons

Flashbang grenades, also called stun grenades, are explosive devices that emit an extremely loud bang and bright lights when detonated, and are intended to disorient people.

## A promise to a friend



Photo: iStock

In a first, researchers use base editing to correct recessive genetic deafness and restore partial hearing.

By Caitlin McDermott-Murphy writing for the Harvard Gazette

When Wei Hsi “Ariel” Yeh was an undergraduate, one of her close friends went from normal hearing to complete deafness in one month. He was 29 years old. Doctors didn’t know why then and still don’t. Frustrated and fearful for her friend, Yeh dedicated her research in chemistry to solving some of the vast genetic mysteries behind hearing loss.

Technologies like hearing aids and cochlear implants can amplify sound but can’t correct the problem. Perhaps gene editing could, scientists decided, since genetic anomalies contribute to half of all cases.

Two years ago, Yeh and David R. Liu repaired a dominant mutation and prevented hearing loss in a mouse model for the first time. But, Liu said, “Most genetic diseases are not caused by dominant mutations. They’re caused by recessive ones, including most genetic hearing losses.”

Now, Liu, Yeh, and researchers have achieved another first: They restored partial hearing to mice with a recessive mutation in the gene TMC1 that causes complete deafness, the first successful example of genome editing to fix a recessive disease-causing mutation.

Dominant disease mutations, meaning those that affect just one of the body’s two copies of a gene, in some ways are easier to attack. Knock out the bad copy, and the good one can come to the rescue. “But for recessive diseases,” Liu said, “you can’t do that. By definition, the recessive allele means that you have two bad copies. So, you can’t just destroy the bad copy.” You have to fix one or both.

To hear, animals rely on hair cells in the inner ear, which ripple under the pressure of sound waves and send electrical impulses to the brain. The recessive mutation to TMC1 that Liu and Yeh hoped to correct causes rapid deterioration of those hair cells, leading to profound deafness in mice at just 4 weeks of age.

Jeffrey Holt at the Harvard Medical School and an author of the paper, successfully treated TMC1-related deafness with gene therapy by situating cells with healthy versions of the gene among the unhealthy to counteract the disease-causing mutation. But Volha “Olga” Shubina-Aleinik, a postdoctoral fellow in the Holt lab, said gene therapy may have a limited duration. “That is why we need more advanced techniques, such as gene editing, which may last a lifetime.”

Yeh spent years designing a base editor that could find and erase the disease-causing mutation and replace it with the correct DNA code. But even after she demonstrated good results in vitro, there was a problem: Base editors are too large to fit in the traditional delivery vehicle, adeno-associated virus, or AAV. To solve this problem, the team split the base editor in half, sending each piece in with its own viral vehicle. Once inside, both viruses needed to infect the same cells so the two base editor halves could rejoin and head off to find their target. Despite the labyrinthine entry, the editor proved to be efficient, causing only a minimum of undesired deletions or insertions.



David Liu is conducting research with Wei Hsi “Ariel” Yeh on gene editing.

“We saw very little evidence of off-target editing,” David Liu said. “And we noticed that the edited animals had much-preserved hair-cell morphology and signal transduction, meaning the hair cells, the critical cells that convert sound waves to neuronal signals, appeared more normal and behaved more normally.”

Of course, more work needs to be done before the treatment can be used in humans. Unedited cells continued to die, causing deafness to return even after the base editor restored function to others. But the study also proved that the clandestine AAV delivery method works. Already, Liu is using AAV to tackle other genetic diseases, including progeria (premature aging), sickle cell anemia, and degenerative motor diseases. “We’re actually going after quite a few genetic diseases now, including some prominent ones that have caused a lot of suffering and energized pretty passionate communities of patients and patient families to do anything to find a treatment,” Liu said. “For progeria, there’s no cure. The best treatments extend a child’s average lifespan from about 14 to 14.5 years.”

For Yeh, whose friend is still living with hearing loss, genetic deafness remains her primary target. “There’s still a lot to explore,” she said. “There’s so much unknown.”

From the [Harvard Gazette](#)

## Free aged care sign language interpreting service

The Australian Government has launched a free national sign language interpreting service for senior Australians who communicate in Auslan and receive or want to access aged care services.

Minister for Aged Care Richard Colbeck announced, "It will ensure that these people can participate in the assessment, planning, and review of their care – something which may have been more difficult in the past."



Certified interpreters will be able to provide Auslan, Aboriginal and Torres Strait Islander languages, American Sign Language, International Sign Language, signed English for deaf or hard of hearing consumers, tactile signing and hand-over-hand for deafblind users.

Both face-to-face and online interpreting, including on-demand video remote interpreting, will be on offer nationally, seven days a week and after business hours when required.

The service supports people to engage with:

- Aged Care System Navigators
- My Aged Care
- Regional Assessment Services
- Aged Care Assessment Teams
- Home Care Packages providers
- Commonwealth Home Support Program providers
- Residential aged care service providers
- Others involved in the provision of Australian Government funded aged care services

All aged care providers and assessors can access the free service. But they will first need to register to get a unique booking code.

More information for individuals and provider or assessor organisations on the [Deafness Forum website](#)

## How Do 'Acoustic Weapons' Work?



Photo: Shutterstock

In recent weeks, acoustic weapons like flash bang grenades and sound cannons have come to new prominence in the public consciousness thanks to their inclusion in the arsenal of weapons used by U.S. police against protesters.

Flashbang grenades, also called stun grenades, are explosive devices that emit an extremely loud bang and bright lights when detonated, and are intended to disorient people. A long range acoustic device (LRAD), also called a "sound cannon," is a speaker system and sound energy weapon is used to disperse crowds by emitting an extremely loud, high frequency noise that can cause pain, disorientation and injury to those exposed.

And although both types of devices are intended only to disorient people, flashbang grenades and sound cannons can cause very serious injuries.

One potential injury from a flash bang grenade is hearing loss. Their detonation causes a noise that is louder than a jet engine and can cause temporary deafness, ear ringing and loss of balance. If a person is near enough to the flashbang grenade when it detonates, the hearing loss may be permanent. Other injuries can result, whether from the force of the explosion or the heat generated by the detonation itself—when the flash powder ignites, it does so with temperatures hotter than lava.

If you're in a position where they are being used against you, there's not much you can do to safeguard yourself from the worst of their effects. The most you can do is to try and avoid getting hit in the first place—which, given the apparent predilection for police use of retaliatory acts against peaceful protesters, is easier said than done.

### **Sound cannons**

A sound cannon looks like a big, bulky stereo system, and is capable of releasing sounds meant to disorient. Injuries from the use of sound cannons can include a persistent ringing in the ears that can last for minutes or days, headaches, nausea, sweating, vertigo and loss of balance. More serious effects can include permanent hearing loss.

Rachel Fairbank writing for [Lifehacker](#)



## **Member of the Order of Australia - Marie Kormendy**

Marie Kormendy was just four-years-old when she knew she wanted to be a teacher, which led her down a path to eventually open WA's first school for deaf children.

"When I was in teacher's college in Ballarat the principal of the newly established training centre for teachers of the deaf in Melbourne came to the college and talked to us about teaching deaf children," the 84-year-old said. "I was so inspired by him that I decided right there on the spot that I wanted to be a teacher of the deaf."

In 1959 Ms Kormendy moved to WA and, some years later, started and became the founding principal of the Speech and Hearing Centre.

"There was very little in terms of options for deaf children [in WA] at the time. When I see the Speech and Hearing Centre now, well I should say Telethon Speech and Hearing, and visit there I'm quite staggered to think that we started it off in the Nedlands Park Tennis Club some 50 more years ago."

Ms Kormendy said the centre started off with just five children, with herself and parent helpers.

She said watching parents realise their children could comprehend things was what kept her going. "I've seen that kind of thing so many times with parents, particularly mothers, of young deaf children when they first see that their child does understand something - it's very precious," she said.

Ms Kormendy has been appointed a Member of the Order of Australia for her "significant service to people who are deaf or hard of hearing through humanitarian assistance programs". She says receiving the award for her work with HearingAID-East Java – which works to provide deaf Indonesian children with access to the latest practices is an honour she must share.

"I'm only part of the journey that's been taken in East Java, so I'm just part of a bigger picture, in a way I feel by accepting it, it's also a tribute to them as well," she said.

By Briana Shepherd for [ABC Perth](#)

## The National Relay Service App

As COVID-19 restrictions begin to ease, remember that you can access some of your favourite National Relay Service (NRS) call options on the go with the NRS app.

The app is free and available on Apple and Android devices.

Using the app, you can make the following calls through the NRS:

**NRS Chat** – if you like to type and read messages

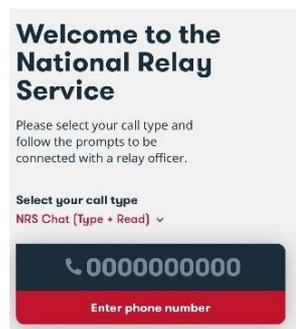
**NRS Captions** – if you like to speak your side of the conversation and read the other person's responses on the screen of your phone or tablet

**Voice Relay** – if people find that you're a bit hard to understand over the phone, a relay officer can re-speak your words to the other person

**Video Relay** – if you want to sign your side of the conversation in Auslan over a video call using Skype and a Relay Officer to translate your signing

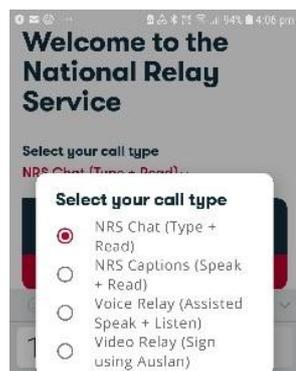
Get the app from the [Apple App Store](#) or [Google Play Store](#).

You can choose which type of call to make on the first screen when you open the app:



If you want to choose an option that's not NRS Chat, tap the arrow

and the full list of options will appear



Just tap the option you want to use and follow the prompts.

### Where do I go for more information?

More information about using NRS Chat, NRS Captions, Voice Relay and Video Relay is available on the [Service Features](#) page of Accesshub.

The [NRS Helpdesk](#) is open Monday to Friday from 8am to 6pm (AEST, excluding national public holidays). You can also leave a message outside these hours and a Helpdesk staff member will get back to you.

Phone: 1800 555 660, TTY: 1800 555 630, SMS: 0416 001 350

Online: [Online form](#) Email: [helpdesk@relayservice.com.au](mailto:helpdesk@relayservice.com.au)

## Deafness isn't a 'threat' to health. Ableism is



Illustration by Brittany England

A few weeks ago, while in my office between lectures, a colleague appeared at my door. We'd never met before, and I no longer remember why she'd come, but in any case, once she saw the note on my door that informs visitors that I'm Deaf our conversation took a sharp detour.

"I have a deaf in-law!" said the stranger as I let her in. Sometimes, I dream up retorts to this kind of statement: *Wow! Amazing! I have a blonde cousin!* But usually I try to stay pleasant, say something noncommittal like "that's nice."

"He has two kids," said the stranger. "They're fine, though! They can hear."

I dug my fingernails into my palm as I contemplated the stranger's proclamation, her belief that her relative — and that I — were not fine. Later, as if realizing this might have been offensive, she backtracked to compliment me on "how well I spoke."

When she finally left me — seething, embarrassed, and about to be late for my next class — I thought about what it meant to be 'fine.'

Of course, I'm used to these kinds of insults.

People who have no experience with deafness often are the ones who feel freest to express their opinions about it: they tell me they'd die without music, or share the myriad ways they associate deafness with being unintelligent, ill, uneducated, poor, or unattractive.

But just because it happens a lot doesn't mean it doesn't hurt. And that day, it left me wondering how a well-educated fellow professor might come to have such a narrow understanding of the human experience.

Media depictions of deafness certainly don't help. The New York Times published a panic-inducing [article](#) just last year, attributing numerous physical, mental, and even economic problems brought on by hearing loss.

My apparent fate as a Deaf person? Depression, dementia, above-average ER visits and hospitalisations, and higher medical bills — all to be suffered by the deaf and hard-of-hearing.

The problem is, presenting these issues as inextricable from being deaf or hard-of-hearing is a gross misunderstanding of both deafness and of the American healthcare system

Conflating correlation with causality fuels shame and worry, and fails to address the roots of the problems, inevitably leading patients and healthcare providers away from the most effective solutions.

As an example, deafness and conditions like depression and dementia can be linked, but the assumption that it's caused by deafness is misleading at best.

Imagine an elderly person who has grown up hearing and now finds herself confused in conversation with family and friends. She can probably hear speech but not understand it — things are unclear, especially if there's background noise like in a restaurant.

This is frustrating for both her and her friends, who constantly have to repeat themselves. As a result, the person begins to withdraw from social engagements. She feels isolated and depressed, and less human interaction means less mental exercise.

This scenario could certainly speed up the onset of dementia. But there are also many Deaf people who don't have this experience at all, giving us insight into what actually allows Deaf people to thrive.

The American Deaf community — those of us who use ASL and identify culturally with Deafness — is an extremely socially-oriented group. (We use the capital D to mark the cultural distinction.)

These strong interpersonal ties help us navigate the threat of depression and anxiety caused by isolation from our non-signing family.

Cognitively, studies show those fluent in a signed language have faster peripheral vision reaction times and directional movement discrimination. Many Deaf people are bilingual — in ASL and English, for example. We reap all the cognitive benefits of bilingualism in any two languages, including protection against Alzheimer's-related dementia.

To say deafness, rather than ableism, is truly a threat to one's well-being, is simply not reflective of the experiences of Deaf people. But, of course, you'd have to speak with Deaf people (and truly listen) to understand that.

It's time to look at the systemic issues that impact our well-being and quality of life — rather than assuming deafness itself is the problem.

Issues like higher healthcare costs and our number of ER visits, when taken out of context, place the blame where it simply doesn't belong.

Our current institutions render general care and technology like hearing aids inaccessible to many. Rampant employment discrimination means many d/Deaf people have substandard health insurance, though even well-reputed insurance coverage often won't cover hearing aids. Those who do get aids must pay thousands of dollars out of pocket — hence our higher healthcare costs.

Deaf people's above-average visits to the ER are also no surprise when compared to any marginalised population. Disparities in American healthcare based on race, class, gender, and ability are well-documented, as are doctors' implicit biases.

Deaf people, and especially those at the intersection of these identities, face these barriers at all levels of healthcare access.

When a person's hearing loss isn't treated, or when providers fail to communicate effectively with us, confusion and misdiagnoses occur. And hospitals are notorious for not providing ASL interpreters though they're required to by law.

Those elderly deaf and hard-of-hearing patients who *do* know about their hearing loss may not know how to advocate for an interpreter, live-captioner, or FM system.

Meanwhile, for culturally Deaf people, seeking medical attention often means wasting time defending our identity. When I go to the doctor, no matter what for, physicians, gynecologists, even dentists want to discuss my deafness rather than the reason for my visit.

It's unsurprising, then, that d/Deaf and hard-of-hearing people report a higher level of mistrust in healthcare providers. This, combined with the economic factors, means many of us avoid going at all, end up in the ER only when symptoms become life-threatening, and endure repeated hospitalisations because doctors don't listen to us.

But, like discrimination against all marginalised patients, ensuring truly equitable access to healthcare would mean more than working at an individual level — for patients or providers.

Because while isolation for *all* people, deaf or hearing, can lead to depression and dementia in the elderly, it's not a problem inherently worsened by deafness. Rather, it's exacerbated by a system that isolates d/Deaf people.

That's why ensuring our community can stay connected and communicate is so important.

Rather than tell those with hearing loss that they're doomed to a life of loneliness and mental atrophy, we should be encouraging them to reach out to the Deaf community, and teaching hearing communities to prioritise accessibility.

For the late-deafened, this means providing hearing screenings and assistive technology like hearing aids, and facilitating communication with closed captions and community ASL classes.

If society stopped isolating elderly deaf and hard-of-hearing people, they'd be less isolated.

Maybe we can start by redefining what it means to be "fine," and considering that the systems abled people have created — not deafness itself — are at the root of these issues.

The problem isn't that we d/Deaf people can't hear. It's that doctors and communities don't listen to us.

Real education — for everyone — about the discriminatory nature of our institutions, and about what it means to be d/Deaf, is our best chance at lasting solutions.



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## Australia won't be 'whole' until Indigenous gap is closed



Minister for Health Greg Hunt. Source: AAP

The Morrison government is investing \$35 million into 42 national Indigenous health projects, Health Minister Greg Hunt says.

These projects include ending avoidable indigenous deafness, ending avoidable indigenous blindness and help to eradicate chronic kidney disease.

Australia's indigenous community is disadvantaged in every social indicator that you can name. Indigenous people with disability are among the most disadvantaged people in Australia. A high incidence of hearing loss is linked to Aboriginal people being over-represented in the prison system. Deafness Forum has advocated that addressing the ear and hearing issues among First Nations people is a key way to Close The Gap.

### Disability in the Bush



The new [Disability in the Bush app](#) uses plain language and culturally relevant information to empower people with disability in remote Indigenous communities to understand the NDIS and make their own decisions. The app explains who is eligible for the NDIS and what supports they can access.

This free app is available in English, Arrernte, and Pitjantjatjara. It was funded by the NDIS.

## Ideal Accessible Communications

Ideal Accessible Communications in Australia is a document that can be used by organisations and individuals working to create an accessible communications sector in Australia.

An ideal accessible communications sector is one in which there are no communication barriers.

The document, prepared by Australian Communications Consumer Action Network brings together the initial insights of 35 Disabled Peoples Organisations, advocacy groups and disability service providers about accessible communications.

Find it on the Australian Communications Consumer Action Network [website](#)

## Deafblind Awareness Week

Deafblind Awareness Week, 22 to 27 June 2020 will feature inspiring videos of deafblind people. There will be no community event this year due to current restrictions on number of people being together. Celebrate the deafblind community around Australia on social media: click on this Facebook [link](#) to register for the event.

## Having trouble with your phone or internet connection?

Australian Communications Consumer Action Network has made a library of free tips for dealing with your telecommunications service provider. Here's where you get the [free resources](#)

## Get help to deal with Royal Commission into Disability

Blue Knot Foundation offers free specialist counselling support and a referral service for people affected by the Royal Commission. Their counsellors have a deep understanding of complex trauma and how it impacts a person.

Contact Blue Knot Foundation National Counselling and Referral Service on 1800 421 46 between 9am and 6pm Monday – Friday and 9am and 5pm Saturday, Sunday and public holidays.

For other support services, visit [www.disability.royalcommission.gov.au](http://www.disability.royalcommission.gov.au)

## Know someone who might like to get their own One in Six?

Drop us a line: [hello@deafnessforum.org.au](mailto:hello@deafnessforum.org.au)

We acknowledge the traditional owners of country throughout Australia, and their continuing connection to land, sea and community. We pay respect to them and their cultures, and to elders past, present and future. We acknowledge the challenge of overcoming high levels of ear health issues among First Nation people and its role in Closing the Gap. We acknowledge the risk to indigenous sign languages of disappearing and the importance of Auslan.

People with disability have and continue to be subjected to isolation, exploitation, violence and abuse in institutions. We thank the Australian Parliament for its bipartisan support of a Royal Commission into the evil committed on people with disability.

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