

mndnews

NEWSLETTER OF THE MOTOR NEURONE DISEASE ASSOCIATION OF NEW ZEALAND INC

www.mnda.org.nz

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Motor Neurone Disease

Motor Neurone Disease (MND) is a condition affecting the nerves that control muscle movement resulting in weakness and paralysis. MND is also known as ALS and Lou Gehrig's Disease. There are various subtypes including Primary Muscular Sclerosis (PMA) and Primary Lateral Sclerosis (PLS). Individuals present with different patterns of symptoms and rates of progression.

The MND Association of New Zealand Inc. exists to provide support, information and advocacy for those living with a diagnosis of Motor Neurone Disease.

Disclaimer: The content of this newsletter is provided for information sharing purposes only. Whilst all care is taken to use only reputable sources, content should not be construed as an endorsement by the MND Association NZ Inc. nor as a recommendation for any individual. We strongly advise you discuss options with those who know you best before making any changes to your routines.

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MND Registry Ready to Launch

A Registry of people with MND in New Zealand will be officially launched in May, sponsored by MND New Zealand. Funds raised for research by the Walk 2 D'Feet MND in 2015 and 2016 are being used to establish and maintain the Registry.

THE NEW ZEALAND MND REGISTRY will connect people with MND to researchers, informing participants if they are a suitable candidate for research studies.

Over time, the MND Registry will facilitate the growth of the MND research field in New Zealand by enabling researchers to find study participants quickly and easily. The MND Registry will also provide valuable information to guide the future development of support services.

It is estimated that over 300 people are living with MND at any one time in New Zealand. We hope that a large percentage of people living with MND will choose to enrol with the Registry. The Registry will collect demographic, contact and clinical

NEW ZEALAND
MND
REGISTRY

information, as well as genetic information if this is known.

Data will be stored anonymously in a secure online database maintained by the Australian MND Registry.

"In rare diseases every patient counts, and the inclusion of New Zealand is a welcomed and vital addition to the MND registry collaboration led by the Australian MND Registry," says Associate Professor Paul Talman, Principal Investigator of the Australian MND Registry.

As well as enrolling people with MND, the MND Registry will also enrol people without MND who have a family member with an identified genetic form of MND (including those whose affected family members have had only fronto-temporal dementia symptoms, if genetic testing has identified an expansion in the C9orf72 gene).

Healthy volunteers needed too

Researchers at the Centre for Brain Research (CBR) at the University of Auckland are working hard to find and develop new treatments for brain disease. If you are interested in contributing to current and future research, you can join the Research Volunteer Register. It requires healthy volunteers as well as people with neurological conditions to participate. More research is essential – and volunteers are too.

Email cbrvolunteer@auckland.ac.nz to join the Research Volunteer Register or request an information leaflet.



Kerry Walker

For more information, see www.mnda.org.nz/registry or contact Kerry Walker, NZ MND Registry Curator: mndregistry@adhb.govt.nz
Ph 0800 MND REG (0800 663 734)



Fancy a Walk?

12th November
Walk 2 D'Feet MND 2017

FOLLOWING THE AMAZING success of last year's Walk 2 D'Feet MND, which raised \$200,000 for MND research and support, we are starting to make plans for Walk 2 D'Feet MND 2017.

This year we have moved the walk date from September to Sunday 12th November – hopefully the weather will be warmer and drier than last year!

We would like even more towns and cities to be involved, so that as many of you as possible can join us on the day.

As always, we can't make these things happen without your help. If you are interested in helping organise a Walk 2 D'Feet MND in your area please contact Claire Reilly at fundraise@mnda.org.nz. Claire will provide you with all the information you need.

We also need volunteers during the lead-up to the walks and on the day, so if you would like to help in your area please do get in touch!

Keep an eye on our website (www.mnda.org.nz) and Facebook page (www.facebook.com/mndanz) for all the latest walk updates.

Sponsors Wanted

WE ARE LOOKING for sponsorship partners for the Walk 2 D'Feet MND 2017. Download the Partnership Proposal from mnda.org.nz/help-us/walk-to-dfeet-mnd and ask your workplace to support these positive and powerful nationwide events that strengthen communities and families, empower people who are facing a tremendous challenge, and create hope.

Nominate your Shining Light

THE DAVID OLIVER BEACON AWARD honours, acknowledges and celebrates those who work with people with MND. It is open to individuals, groups and services that provide a positive contribution, improvement or innovation in MND support or care. The theme of this year's Award is 'Shining Light'.

We welcome nominations from people with MND, their carers and from healthcare professionals. We would love to know who your 'shining lights' are and what makes them exceptional in their field.



Download the nomination form from www.mnda.org.nz. Nominations close on June 21st.

The 2016 David Oliver Beacon Award winner was Karen Thomas, the neurology nurse specialist based at Waikato Hospital.

Goodbye, Cornflowers Remembrance Field

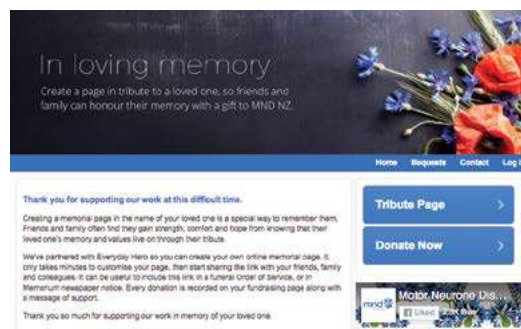
WHEN THE NEW WEBSITE for MND New Zealand was launched last year, we had to bid farewell to the MND Cornflowers Remembrance Field from our previous website. Many families had dedicated cornflowers and memorial messages to the memory of loved ones on this graphic element of our website, and we are very sorry that we were unable to transfer these tributes.

Unfortunately, the technology used to create the Cornflower Field is no longer supported and now poses a security risk. The cost to redevelop the Cornflower Field in a safer format for our new website was prohibitive.

Families who wish to donate to MND New Zealand while paying tribute to a loved one can now create an 'in memory' page on the Everyday Hero website: www.everydayhero.co.nz/event/InmemoryMNDNZ

Tribute pages can be customised with a photo of your loved one, and the link is easily shared with friends, family and colleagues. It can be useful to include this link in a funeral Order of Service or newspaper notice. Every donation is recorded on the tribute page along with a message of support.

Thank you for supporting our work in your time of loss, and for your generous decision to contribute to the care and support for others with MND.





Farewell to our Fieldworkers

NO, WE ARE NOT LOSING ALL OUR STAFF – but we are delighted to announce a change to their job title!

For many years we have been uncomfortable with the job title Fieldworker,

which evolved from the post-war title of Field Officer. While the second part of the name does describe well the incredible effort our team put in, the first part of the title suggests work on a farm, so we have agreed it is time for a change.

It is a challenge to describe just what our Fieldwork Service provides, because it varies from setting to setting, person to person, DHB to DHB, community to community – and we believe that is our strength. Every time we try to pin the role down, it comes back to **support**.

By its very nature, the support we give varies across individuals. Some would ideally like someone to pop in every week for a cuppa, others prefer we keep our distance until called upon. Early after diagnosis it is common to want little involvement with us, but over time we hope to build a relationship with people, so that when you don't know who to call – you call us!

Thanks to the fundraising efforts of so many, we are able to say “I can support you in whatever way suits you”. Although we can't provide funds or equipment, we are always the on the end of the phone, email or text and will respond as quickly as we can. Our team are all employed part-time but have back-up from National Office.

After much deliberation, in future we will refer to our community team as **MND Support Workers** who provide our **MND Support Service**.

Business carries on as usual! Do contact our MND Support Team if you need support. Tell us what support means to you, and we will do our best to deliver.

Your Feedback

We were very happily surprised by the responses when we announced the change to the health professionals in our database, and greatly appreciated their feedback:

“I think the change of name is a very good idea as they do provide a lot of support to patients, families and other healthcare practitioners too.”

“Sounds good, we value our Support Worker immensely – a great team leader.”

“Can we please pass on our recognition that our local MND Support Worker is an amazing resource to both us and our families, and we much appreciate the work she does.”

“It is hard to find a title that does the role justice. Your team works so hard and is very much part of the interdisciplinary team. Our work would be harder without you and I am sure we would miss the vital support and information gathering that you carry out.”

“You do a great job of supporting our MND patients in this area – we really do appreciate it.”

Our MND Support Team

We employ six part-time Support Workers around the country. Please contact your closest Support Worker and they will respond as promptly as possible. If you live some distance from your Support Worker, contact will mainly be by phone, email and text, with occasional visits. We do not provide out-of-hours or emergency services, or medical advice.

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Genetics and MND

Everything you need to know

There has been a rise in peoples' interest in the genetics of MND in recent years, thanks to research developments in this area. **Courtney Forgeng**, a certified genetic counsellor, shares some information on MND genetics and the services offered by the New Zealand Genetic Health Service.



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THE MOST IMPORTANT THING for families to know is that most motor neurone disease (MND) is sporadic. This means that it does not run in families. Only ten percent of MND is estimated to be hereditary.

The New Zealand Genetic Health Service sees people where a mutation is already identified in their family, or to assess a family that has not had any testing.

Testing needs to start in a family member with a clinical diagnosis of MND or dementia, as this gives the most information for a family.

A family has to have two or more first-degree (eg parent or sibling) or second-degree (eg grandparent, aunt, uncle) relatives with MND and/or early onset dementia to qualify for an assessment. All appointments are funded for New Zealand residents who meet referral criteria. A referral can come from a GP or specialist.

During the first appointment an exten-

sive family tree is taken, and the implications of testing are thoroughly discussed. Assessment of the family history can inform how MND is being inherited in the family, for example in a dominant fashion, where all children have a 50% risk of inheriting the gene.

Genetic testing only identifies the cause in around 30% of familial MND. This means for many families the genetic cause remains unknown. However additional tests may be available in the future.

Some people prefer to store their DNA for later use by their families instead of undergoing genetic testing.

Pre-Symptomatic Testing

When a mutation is known in a family, at-risk relatives can make an appointment for pre-symptomatic testing. The testing process may slightly differ at each Genetic Health Service Hub (see box); however pre-symptomatic testing usually requires several appointments. During

the appointments we discuss family history, the inheritance pattern, the person's motivation for testing, and the effects of testing on relationships. We later meet in person to review the results.

This protocol gives time for the person to process the information, think about the pros and cons, and to be able to best adapt to the result. Some people stop the process after the first or second appointment as they decide they do not want to know, or want to delay knowing their result.

As there is no cure for MND, many people do not wish to know their status. The Genetic Health Service supports both the right to know and the right not to know. Others feel they need to know their result to properly plan for the future and make the most of their time.

The timing of the test can be very important. Two of most common times people seek genetic testing is before they have children, or before they have grandchild-

dren. The Genetic Health Service does not test people under 18 years old.

Family Planning

Many families would like to know the chance of passing MND on to a future generation, and some would like to undergo measures to prevent passing on MND. The New Zealand government provides funding for family planning options such as testing during a pregnancy, or with IVF pregnancy, testing embryos known as Preimplantation Genetic Diagnosis (PGD). Both these testing options can only be done if a family's genetic mutation is known.

A separate appointment with the Genetic Health Service is required to thoroughly discuss family planning options and the PGD process. There is a substantial waitlist to access PGD at present. This will be discussed when the couple is seen by a fertility clinic.

Insurance

If you chose to undergo genetic testing, there may be an impact on your ability to obtain new insurance policies. You would have to declare genetic test results or family history if asked, and it may mean that you would not be covered for anything to do with MND and that your premiums become higher. This may not be an issue for you if you have insurance in place already. It may be advisable to speak to an independent financial or insurance advisor.

The Genetic Health Service comprises three Hubs:

Northern Hub covers the top of the North Island down to Taupo and Gisborne

Phone (09) 307 4949 (ext 25870) or 0800 476 123

Email: GenSec@adhb.govt.nz

Central Hub covers the rest of the North Island and Nelson

Phone (04) 385 5310 or 0508 364 436

Email: genetic.services@ccdhb.org.nz

South Island Hub covers the rest of the South Island

Phone (03) 378 6574 or 0508 364 436

Email: genetic.servicenz@cdhb.health.nz

Light a Candle

WE LOVE the beautifully presented and divinely scented candles made by Cienna Boutique (www.cienna.co.nz). All candles are handmade in New Zealand with 100% soy wax.

With Mothers' Day just around the corner, why not "light up your Mum's life" with a candle, or treat yourself so you can light a candle in remembrance if you've lost your mother.

Enter the letters MND in the Order Notes box when you complete your order, and the kind owner Stacey, whose mother-in-law has MND, will donate 20% of the total sale price (excluding P&P) to MND NZ.



MND? There's an App for That

A NEW SMARTPHONE APP, the Mobile ALS Analyzer, aims to create the world's largest source of information about the progression of MND. It has the support of the International Alliance of MND/ALS Associations, which includes MND New Zealand, and we are encouraging our clients to give it a try if they have a smartphone.

The simple app asks users to frequently complete simple tasks, and uses feedback from the phone itself to measure daily activity.

By collecting this objective, continuous data, the app can assess the functional abilities of people with MND and the progression of the disease. The developers hope to create the world's largest database of MND clinical manifestations, and intend to analyse the information to seek out behavior correlations using advanced algorithms.

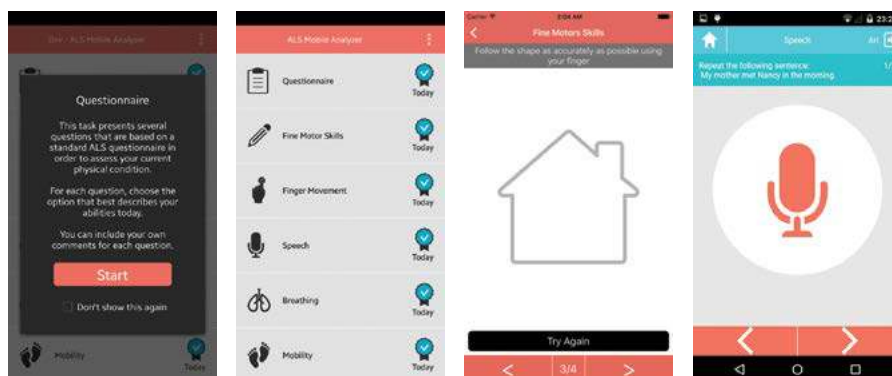
Eventually this will improve clinical care for people with MND, by enabling doctors to get more accurate and frequent clinical data.

It will also make future clinical trials millions of dollars cheaper, by simplifying participant monitoring, shortening trial times, and enabling more frequent, accurate and objective assessment of treatment efficacy, which is the most important goal of a clinical trial.

Prize4Life initiated and developed the Mobile ALS Analyzer app, with the project directed by Shay Rishoni, who has MND.

The app is free for people living with MND anywhere in the world, and also those who wish to use the app as healthy controls in order to provide a "healthy" baseline. Sign up if you're willing to use the app frequently, because the more often data is collected, the more useful the results.

For more information, check out the website (www.alsanalyzer.com) and watch the 15 minute presentation from the recent International ALS/MND Symposium (www.youtube.com/watch?v=2mOhQhwkNfI).



There's a Hole in my Bucket

When Mike Baker, the great-grandson of author JRR Tolkien, died due to MND aged just 39, he left his brother Royd Tolkien a bucket list of 50 challenges to undertake in Mike's memory. Royd has been in New Zealand this year, making *There's a Hole in my Bucket*, a documentary about completing his brother's bucket list. This is Royd's story.

EVERY FAMILY HAS ONE, the kid with an inborn, insatiable appetite for risk, always on the lookout for the next rush. That's my little brother, Mike.

Even when he was young he'd be jumping his bike as far as he could or climbing to the top of every tree while I watched horrified. As he got older, he got bolder. He loved any form of physical activity and was always unsuccessfully trying to drag me along, be it jumping out of a plane, snowboarding, skating, knee-boarding – basically anything that got his heart rate up.

In September 2012, Mike was diagnosed with motor neurone disease. On January 28th 2015 my beautiful, strong, fearless brother passed away.

When we found out his diagnosis, we went to a very dark place. As his big brother I had a duty to look after and protect him, and being told you are absolutely powerless to help as your brother becomes very ill, very quickly, was hard to come to terms with and difficult to accept. But however difficult for me, it was nothing compared to what Mike dealt with daily.

MND is devastating for anyone, and a particularly cruel hand to be dealt to an active adrenalin junkie like Mike. The



illness gripped him tight, but we linked arms and faced it together right until the last moment. Together with his girlfriend, I spent the last 3 years caring for him, and while it destroyed me seeing him little by little lose the ability to do anything, he constantly blew my mind with his strength, spirit and determination.

Mike always wanted to go to Queens-town, the adventure capital of the world, where he could feast on crazy adrenalin sports, but he never managed to fulfill his dream. So he set me a challenge: to do it for him. He drew up a seriously badass bucket list of 50 items, and instructed me to tick it off by any means possible and face my fears as he faced his.

This was no ordinary bucket list. Mike loved orchestrating awkward embarrassing situations that would make me look

as big a buffoon as possible. There was no greater joy to me than to see him laugh, and he laughed the most when he could set me up for a fall!

I'll admit I'm a wuss and haven't a thrill seeking bone in my body. I prefer cups of tea and gardening and I certainly do all I can to avoid tricky situations. But my lily-liver has taken a back seat as I've been completing Mike's insane list of thrill-seeking tomfoolery. I'm experi-

encing a part of his life that he loved so much while knowing that he'd be splitting his sides at the compromised situations he's put me in!

I'm doing this because I've learnt some very important lessons over the last few years. I've seen my brother go from an incredibly active, healthy man in the prime of his life, to someone whose body gave up on him, needing care 24hrs a day. He taught me to grab life by the balls, never have any regrets, enjoy every breath and always look for the next smile. This gave me a rock solid resolve to do everything he has put on that list.

This project has been more than just a bloke on a jolly doing crazy dares. It is the strength of two brothers' bond built while daring to face his mortality together against a very real ticking clock. It is a journey of discovery and inspiration, about how sweet and precious life can be in the very little time we live it.

My hope for those watching *There's A Hole In My Bucket* is to ignite a passion for life we all should have raging within us and to inspire people to take absolutely nothing for granted. To fully appreciate the family and friends we hold dear and close, and not wait until something terrible happens before realising that. And through all of this, the documentary will also raise the awareness of MND, and show families affected by MND that they are not alone and definitely not forgotten.



Thank you, Royd

While filming the documentary Royd Tolkien raised awareness of MND all over New Zealand, and has been a charming ambassador for MND New Zealand. He very generously helped produce a fundraising event for us in Wellington

on April 12, sourcing some fantastic auction prizes. Thank you so much, Royd! Big thank yous also go to our volunteer event organiser, Jodie O'Doherty, sponsors ResMed and Angel Street, and everyone who attended the event and bid at our auction. We raised over \$15,000!

Sharing the Love

CONGRATULATIONS TO newlyweds Nafeesa and Mike Prestidge, who married earlier this year. Nafeesa and Mike made the very generous decision to request donations to MND New Zealand instead of gifts from their wedding guests.

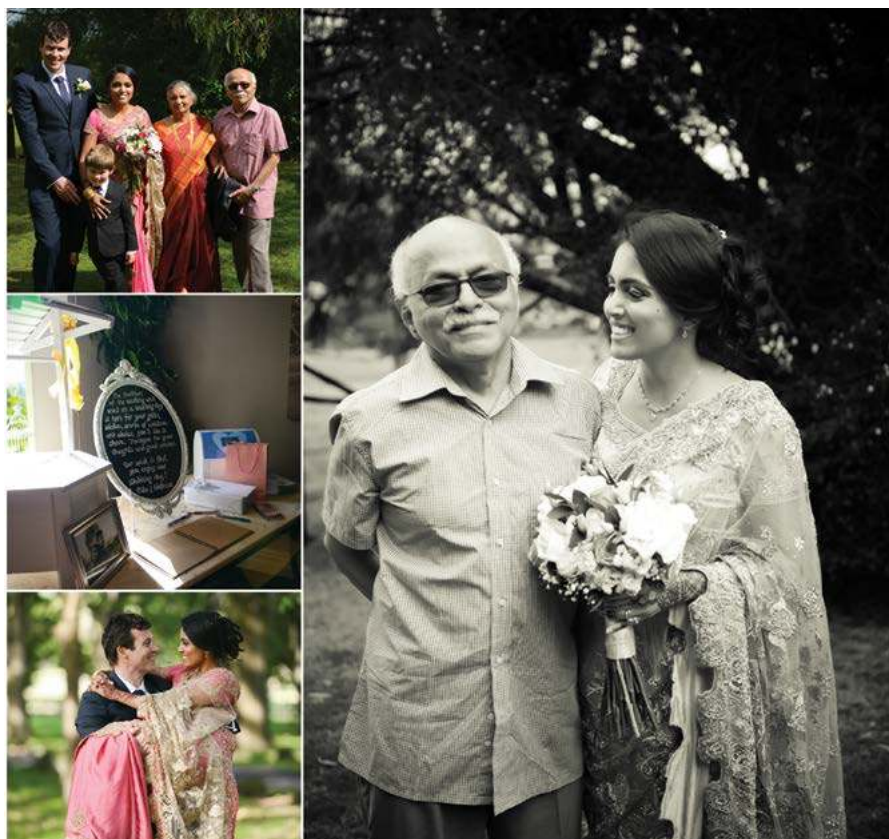
"Mike and I wanted to start our life together by giving something back," says Nafeesa. "My Vappa [Dad] was diagnosed with motor neurone disease last May. We are inspired by the work of MND New Zealand and wanted to support them by raising money to help them continue providing support for people with MND and their caregivers."

Nafeesa says her father's diagnosis

with MND was her worst nightmare. "I am my Vappa's little girl," she says, and her greatest wish for her wedding day was to have her father right beside her. Fortunately her wish came true and "it really was one of the best days in my life so far".

MND is a rough journey for families, Nafeesa says, but the MND community can be a great support. "Since the diagnosis I have met many people with MND through social media. Some became very good friends and will always be close to my heart."

Thank you so much for your support Nafeesa and Mike, and thank you to your very generous wedding guests.

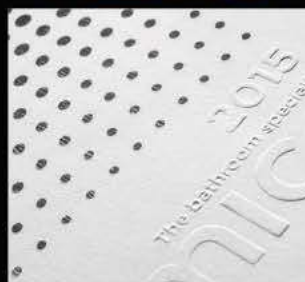


How to fundraise at an event

Nafeesa and Mike created a fundraising page on the Everyday Hero website, and sent the link to guests along with other 'good to know' wedding information, like what time to arrive and directions. They also had a donation box at the wedding reception.

It's easy to create your own fundraising page on Everyday Hero. Go to give.everydayhero.com/nz/get-started, and type MND in the charity search box then select Motor Neurone Disease NZ. Be ready to add your details, a photo of yourself and to write a little about your personal connection to MND to help others connect with your cause. It takes about five minutes to create a page, then you're ready to share it with friends, family and colleagues via email and social media.

Good luck and thank you!



HOUND & STEED
CREATIVE AGENCY

We are proud to have sponsored all the creative needs of MND New Zealand over the past two years and we look forward to working with the great team of passionate people who work tirelessly to make a difference.

MND and a Kiwi Bloke

Graham Jones from the MND NZ Support Team shares the 'number 8 wire' style ingenuity of one of his clients.

MURRAY IS YOUR classic Kiwi man. A builder by trade, he is down to earth and he'll welcome you with a smile. Murray is a man of the land; he has a wealth of knowledge on where to catch a fish, he can identify all of the native trees, and he's a keen hunter. His home is a lifestyle block in Acacia Bay, Taupo.

Four years ago, Murray was diagnosed with MND.

Last week I sent Murray the usual text, as he is non-verbal, asking if it was okay if I dropped by. The usual reply: "I'll put the jug on".

When I arrived, Murray had a glint in his eye. In the garage was a new ride-on mower. Knowing Murray can no longer walk, I wondered exactly how this was going to work. So I asked, "Do we get a demo?"

As can be seen from the photos, Murray together with a mate made some modifications. On the deck of the mower they mounted two handrails, to



enable the transition from chair to chair. The mower is steered by a joystick rather than a steering wheel.

"To be honest, Graham," he tells me, "I finished building, sold the boat and I've not had much to exercise my brain – but this mower has been good for me! The secret is the joystick. I have been a project guy for ever."

I hope Murray's story may provide some inspiration to folk out there. I was speechless watching Murray transfer from his power chair to the mower, then drive off and cut his lawn just like it was any other day. Thank you Murray, you have already made a difference.

Graham Jones



Guide for Carers

The Guide for Carers book has been updated and can be viewed or downloaded from www.msd.govt.nz/what-we-can-do/community/carers/

Thank you, Lynne

It is with regret we advise that Lynne Neshausen has resigned due to ill health. Lynne has provided our clients, and their carers, families and health professionals, with excellent support and service since 2013. Lynne has established strong MND networks in the Taranaki DHB area and has been a respected colleague in the Waikato DHB area. Lynne has been a valuable member of our Support Team and we have appreciated her wise counsel and advice.

Tips, tricks and inspiration



➤ **Thank you** to Steve Green, who makes a regular donation to MND New Zealand from his Auckland business, Steve Green Blinds. Steve has had his company car sign-written with "We support MND New Zealand". Thanks for your awareness raising and kind donations, Steve!



A LOVELY CLIENT, Sandra, is laughing in the face of MND by having a 'Nil By Mouth' T-shirt printed to wear now that she is no longer able to take food and drink orally.



➤ **CONNECTING** a Bluetooth speaker to a tablet computer is a great way to amplify speech produced by an app such as Predictable. The speaker can be placed near the listener for maximum clarity. Jennifer Jenvey recommends the Jawbone model pictured, and says she got it for a good price from Amazon.



➤ **JENNIFER JENVEY** also created a lanyard to inform anyone assisting her that she is unable to speak, but can hear and understand, and includes emergency contact phone numbers.

Please send us your tips, tricks and innovations that help ease life with MND.

Email comms@mnda.org.nz



Australian Research News

Urine biomarker

RESEARCH IN AUSTRALIA has led to the discovery of a new and simple test to monitor disease progression in people with MND, and measure the effectiveness of treatments being tested in clinical trials.

The research, supported by the MND Research Institute of Australia, found that a simple urine test can measure levels of a protein called “p75”. Previously known to be present after nerve injury, the new study found that the amount of p75 was significantly higher in people with MND compared to those without the disease. The more MND progressed, the more p75 was found in the urine.

This suggests that p75 can be used as a “biomarker” that reflects motor neurone degeneration in people with MND. Because urine is easy to collect, this new discovery will facilitate carrying out clinical trials significantly. p75 is currently the only biological-fluid based biomarker of MND progression.



The study, led by Flinders University researchers Dr Mary-Louise Rogers and Dr Stephanie Shephard, and Professor Michael Benatar from the University of Miami, was published in the prestigious journal *Neurology* on 23 February 2017.

New drug discovery

A STUDY LED BY University of Queensland researchers Associate Professor Trent Woodruff and Dr John Lee has found that a drug “PMX205” extends the life of mice that have the SOD1 gene mutation that can cause familial MND. Future studies are planned to test PMX205 in sporadic MND models.

The research, published in the *British Journal of Pharmacology* in March 2017, found PMX205 slowed disease progression and significantly increased the muscle strength of mice. The drug works by blocking a key component of the immune system, reducing inflammation.

More studies are now underway to determine the safety of PMX205 before it can be tested in humans. This will take about two years, after which human testing may occur. The drug has obtained FDA (Food and Drug Administration) and EMA (European Medicines Agency) ‘orphan drug’ approval, which allows for an accelerated progression to human trials.



Trent Woodruff

Copper ATSM progress

IN MARCH, the ALS Therapy Development Institute (ALS TDI) in the US announced that it had replicated findings that Copper ATSM (CuATSM) can slow down MND progression in a mouse model of the disease.

This independently validates the work of Dr Peter Crouch at the University of Melbourne in Australia, who is one of the key researchers working to show the efficacy of CuATSM as a treatment for MND.

This is the first time in 15 years of testing drugs rigorously that ALS TDI has confirmed the efficacy of a proposed treatment for MND that others have previously published.

Dr Dominic Rowe, of Macquarie University in Australia, is the Principal Investigator of a Phase 1 clinical trial that is recruiting participants in Australia to examine the safety of CuATSM in humans.



Dominic Rowe

Stay Up-To-Date with MND Research

ALS News Today

A daily digital news journal that reports the latest science and research news about MND/ALS. alsnewstoday.com

MND Research Blog

Research updates are regularly posted on this blog, run by the UK MND Association. mndresearch.wordpress.com

Clinical Trials Database

A comprehensive database of clinical trials for MND/ALS worldwide. www.alstdi.org/als-research/als-clinical-trials

ALS Research News

The latest research articles and news from ALS.net and ALS-TDI.org. www.als.net/news/research

In The News

The ALS Association website's news section. www.alsa.org/news

Five Promising Treatments

Antisense Drug Breakthrough

'Antisense' drugs are a major breakthrough in MND research, with great promise for the treatment of genetic MND. This field of research received a huge boost just before Christmas, when the US FDA approved the antisense drug Spinraza (nusinersen) for the treatment of spinal muscular atrophy (SMA), a genetic disease that causes the loss of motor neurons in infants and toddlers.

The Phase 3 trials of Spinraza were stopped early after achieving such positive results researchers concluded it was unethical to deny the treatment to the babies in the control group.

MND researchers are enthusiastic about the success of nusinersen. "It is exceptionally exciting," wrote Jeffrey Rothstein of Johns Hopkins Medical Institute, who is developing antisense drugs for the repeat expansions in the C9orf7 gene that can cause MND.



Don W. Cleveland
taken in
2013.

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/ WIKIMEDIA
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"The news is highly uplifting," commented Dr Don Cleveland of the University of California. "It supports the general approach of using designer DNA drugs for changing synthesis of genes in a spectrum of neurodegenerative diseases."

Dr Cleveland, who was not involved in the SMA study, is an antisense pioneer whose lab is collaborating with Ionis Pharmaceuticals to develop antisense drug therapy against mutations in the C9orf72 gene and SOD1 gene, the two leading causes of familial MND, as well as the mutant gene that triggers Huntington's disease.

Antisense technology could reduce the production of SOD1 and C9orf72 proteins in the body, and has the potential to become a therapeutic treatment to slow or even halt MND progression.

A second Phase I study of the SOD1 antisense drug has been underway since January 2016 to test safety in people living with MND. A similar trial targeting C9orf72 is anticipated in 2017 in partnership with Ionis Pharmaceuticals and Biogen.

Dr Cleveland says the SMA study demonstrates that antisense drugs can be effectively delivered broadly across the central nervous system, where they engage their target genes.

"I am convinced that it is time to make a serious effort to treat ALS using gene therapy," says James Wilson, director of the Orphan Disease Center at the University of Pennsylvania, which has launched The Program of Excellence for Motor Neuron Disease.

This program will build on a recent discovery Wilson made regarding gene transfer vehicles, known as vectors. These vectors can transport genes into central nervous system cells and have been used in several clinical trials, including ones for spinal muscular atrophy (SMA). The SMA studies suggest Wilson's technology could be used as a mechanism for treating MND. The program will advance in three phases, beginning with people who have defects in the C9orf72 gene.

NurOwn

Stem cell technology is one pathway being investigated to treat sporadic MND, and the Phase 2 trials of a treatment branded NurOwn have shown positive results.

The Phase 2 trials of NurOwn showed the treatment was safe and well-tolerated, and also showed that some patients' disease progression slowed during the trial period. Four of the 35 people treated in the US trial showed improvement (see tinyurl.com/z88qree for more detail).

NurOwn is not yet available to people with MND outside of clinical trial.

GM6

In January, Genervon Pharmaceuticals announced that its sporadic MND drug candidate GM6 modulates 89 genes that are associated with MND. These genes are involved in pathways that include neuronal generation and regulation of neuron death.

By targeting many genes, GM6 may be a promising therapy approach that tackles different disease-associated path-

ways at the same time.

GM6 was developed to target several neurological disorders, including MND, Parkinson's, Alzheimer's and Huntington's diseases. It aims to improve neuronal repair and survival, and restore their proper function.

It has been shown to have a safe profile in a Phase 1 and three Phase 2 trials with patients with MND, Parkinson's and stroke. According to Genervon, a Phase 3 trial for MND is expected to start this year. (See f1000research.com/posters/5-2836 for more info.)

GM6 is not yet available to people with MND outside the clinical trial.

Masitinib

Another MND drug in the pipeline for approval by the FDA in the US is masitinib, by French company AB Science.

Masitinib is an anti-inflammation treatment that protects muscles and nerves against damage. It is now in a Phase 2/3 clinical trial to find its efficacy in MND. Interim data shows that masitinib, taken along with riluzole, improved patients' quality of life and ability to survive.

Masitinib is an oral treatment that inhibits proteins known as tyrosine kinases. This reduces inflammation in the part of the nervous system that regulates muscles, as well as in the central nervous system.

Masitinib is currently sold in a veterinary formulation for the treatment of mast cell cancer. It is currently approved by the FDA for "compassionate use". Access is allowed in the US only by request of a physician, FDA review and authorisation, and permission from the drug manufacturer.

Edaravone

Edaravone, a product of Japan's Mitsubishi Tanabe Pharma, is also being reviewed by the FDA for approval of use in the United States.

Edaravone (also called Radicut) is approved in Japan for the treatment of stroke and MND. A synthetic free radical scavenger, edaravone works by reducing protein nitration.

Clinical trials are currently being conducted for edaravone in MND patients in Europe. It is not available to people with MND outside Japan.

Taking Control

A Guide to Individualised Funding

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IF Host Providers

National (excluding Auckland): Access Homehealth Ltd, 0800 284 663, www.access.org.nz

National: Healthcare NZ Limited, (06) 8344567, www.healthcarenz.co.nz

National: Manawanui In Charge, 050804620427 (09) 444 8440, info@incharge.org.nz, www.incharge.org.nz

Northland: Home Support North, (09) 401 6657, homesupport@xtra.co.nz, www.homesupport.co.nz

Auckland: Presbyterian Support Northern (Coactive Disability Support Services), (09) 622 4799, info@coactive.org.nz, www.coactive.org.nz

Auckland: Lifewise Homecare Services, (09) 302 5390, lifewise@lifewise.org.nz, www.lifewise.org.nz

Auckland West: Vision West, (09) 818 0700, Office@VisionWest.org.nz, www.visionwest.org.nz

Thames/Waikato: Te Korowai Hauora O Hauraki, (07) 868 5375, www.korowai.co.nz

Tauranga/Bay of Plenty: Whaioranga Trust, (07) 544 9981, reception@wtga.org.nz

Eastern Bay of Plenty: Disability Resource Centre Trust, (07) 307 1447 Freephone: 0800 227 363, www.drct.co.nz

Mid Central/Wellington: Presbyterian Support Central, (04) 439 4900, info@psc.org.nz, <https://central.ps.org.nz/>

Nelson Marlborough/Blenheim: Florence Nightingale Agency, (03) 548 0623, nelson@florence2care.co.nz, www.florence2care.co.nz

Dunedin/Otago: Dunedin Home Support, (03) 455 0714, dunedinhomesupport@xtra.co.nz, www.dunedinhomesupport.co.nz

Where can I find more information?

The Ministry of Health website is a great place to start. Reviewing the IF Host Providers websites and talking to people who use IF is also likely to help decide if IF is right for you.

Individualised Funding is a way of paying for disability related supports that gives you more choice and control.

INDIVIDUALISED FUNDING (IF) is a mechanism for disabled people to have more choice and control over how their disability related supports are provided in their homes and when accessing the community.

When choosing to self-manage personal supports through IF, individuals gain the flexibility to employ who they want to and negotiate times of service that work around their own lives.

IF began as a pilot under the Ministry of Health in 2003, and has since expanded to be available to all disabled people who have been assessed by a Needs Assessment and Service Coordination (NASC) agency, and allocated hours of support for either home help or personal care. This type of support is known as 'home and community based support'.

In practical terms, IF allows an individual to purchase and pay for their home and community based support, with the funds being managed through a third party or IF Host Provider, not the Ministry of Health directly.

Anyone who wants to use IF first needs to meet with a Needs Assessor at their local NASC. The Needs Assessor can refer the individual to an IF Host Provider in their local area, or one of the three National IF Host Providers if preferred.

The amount of funding available to an individual each year is based on a fixed unit price per hour of support allocated. The yearly time frame begins from when your needs assessment and review of supports happens, rather than a calendar or tax year.

The flexible use of funds means that supports can be flexed up or down during the year, depending on when you have the greatest needs. IF must be managed in accordance with Ministry of Health policy and Inland Revenue requirements. At the end of the year, any unspent funds will be returned to the Ministry of Health.

Challenges of IF

Some of the challenges associated with IF include time, record keeping, knowledge, skills, employment relations and personal liability. Many of these issues can be overcome by working with an IF Host Provider who for a small fee will offer coaching, administration tools, and payroll services as an optional extra, for those who prefer not to deal with payroll calculations and tax matters.

You can also nominate another per-

son such as a family member to act as an Agent on your behalf and manage the IF funding arrangements for you.

Individuals who employ support staff through IF (rather than purchasing support services from a provider) carry a personal liability as an employer. This is an important consideration, as things can become costly and stressful if employment relationships break down and a former employee takes up legal action against you. Community Law Offices are

usually inclined to represent employees in such matters, so affordable legal options may be limited. IF providers may be able to inform you about risks associated with this liability and resources they can provide, such as access to an Employers and Manufacturers Association (EMA) subscription.

Thanks to Muscular Dystrophy New Zealand for sharing this article with MND New Zealand.

Making the Most of Individualised Funding

NATALIE BRUNZEL explains why we need to talk about Individualised Funding.

When you type "Individualised Funding" into Google, you will find the Ministry of Health guidelines that provide individuals with an overview of eligibility and the support you can receive. However, if you want to make the most out of IF then you need to discuss it with others who know how the system works. There are a wide range of ways to utilise your hours and once you are in the system you learn that the guidelines are just where the journey begins.

IF works well for those who want to be in charge of their own supports, however it should not be forced onto those who do not wish to manage their own support workers or personal assistants.

A history of IF

In order to understand IF it is important to know how it eventuated. The way in which disability supports are funded has changed and with it we have seen a shift away from institutionalisation towards independent living.

Traditionally, individuals were assessed for their daily needs and then a set amount of hours were granted to support that person. A support agency would provide caregivers up to the allotted hours of care. This process led to organisations having all the power, as the hours were prescribed and the organisations chose

the caregivers, as well as the times at which the individual could receive the support. This led to many people being required to go to bed at 7pm or get up at 10am – not conducive to living an active life within the community! It also meant that if you didn't use the hours at the time given, then you would lose them. They wouldn't automatically accumulate.

IF has been the government's answer to this issue. This shift has seen people re-name their caregivers as personal assistants or support workers. This might seem like a minor shift, but in the psychology of individuals there is a difference between having to care for someone and supporting someone. Being the manager keeps the care more honest.

Not only can individuals using IF choose who to employ as their support people, but they are also able to choose how and when these supports are able to be accessed.

Managing your IF budget

Many of my friends commented on the fact that now they were able to 'stretch the hours' to give them the supports they actually needed. They had observed that people with neuromuscular conditions often didn't receive the hours required to live independently. Therefore, they found ways in which to use IF to support them effectively. One of those ways, for example, was having a family member support them for a week, which then meant they could save up their hours and live indepen-

dently for the rest of the month.

There are many creative ways that individuals have found to make the money stretch further. However, there was an audible panic among my friends when the minimum wage was being raised. Not because they didn't want to pay their support staff more, but rather because it meant further strain to an already tight budget.

Now do not misunderstand, everyone wants to be able to pay their personal assistants as much as possible. However, for some the reality is that they pay the amount they can afford. One of the joys of IF is that disabled people have the choice to pay assistants the amount they decide on. This means that many are paid over minimum wage, because in order to retain good people and get the quality support required, there is a need to increase their pay on a regular basis. But all of this has to be carefully managed.

If you are on IF you are probably aware of many of these things. But, it requires our continued engagement with the Ministry of Health to keep IF honest and functioning in the best possible way. So I urge you all to continue letting your concerns be heard with feedback to MOH and other organisations.

Natalie has worked within the disability sector for the past 10 years and has a passion for ensuring equality.

President's Piece



Kia ora koutou.

Our 2016 Walk 2 D'Feet MND was a major success. Not only did hundreds of people turn out for MND, whatever the weather, but also hundreds of people fundraised for MND, and scores of businesses generously supported us. And overall we raised almost \$200,000 – a result beyond our wildest dreams.

As most of you will know, half of our Walk income is tagged for research. We believe the research initiatives we have chosen to support will build a wider MND community of interest in both the clinical and scientific communities in New Zealand. In turn we believe this will benefit people living with MND now, as well as benefitting future generations through New Zealand's contributions to the fight to find a cure.

One of the exciting initiatives we have funded with money raised by the Walk 2 D'Feet MND in 2015 and 2016 is the New Zealand MND Registry (see page 1).

The other initiative being funded with the money our community has raised is an MND research network or "hub" led by the very talented Aotearoa Fellow Dr Emma Scotter. This hub will provide an infrastructure for connecting existing MND researchers, both clinical and biomedical, in New Zealand. Dr Scotter envisages the hub as "a beacon to attract New Zealand researchers to conduct more MND research".

Dr Scotter, who did post-doctoral research with Professor Chris Shaw at the internationally esteemed King's MND Care and Research Centre in London, has a passion for research into MND. She believes that there are unanswered questions about MND that are specific to New Zealand. She has seen the benefits of a research infrastructure overseas, and is confident that this initiative will lead to more MND research being done in New Zealand. She also foresees the network facilitating interaction between researchers and the public through its proposed website.

Our Walks are very important to MND New Zealand. They bring in all-important income for us to continue to deliver support to people living with MND. They also build our MND community, and help the public understand MND better. And now, with the launch of the New Zealand MND Registry and the MND research network, the Walks are contributing to the growth of knowledge about MND in New Zealand.

These projects would not have happened without your participation, as walkers, as volunteers, as fundraisers, and as donors. Thank you all.

Our 2017 Walk 2 D'Feet MND will be on 12 November (we are hoping for more settled weather). Volunteers are welcome and hugely appreciated. Several other activities are also being planned to raise money and awareness for MND – film-making; a cocktail evening; a night of orchestra. We are inspired by the energy, imagination and diversity of the people making these events happen and we thank each and every one of them most sincerely.

MND is a cruel disease. It is relentless and unforgiving. I hate that no cure has yet been found. It heartens me and gives me hope when people come forward to help us with fundraising and awareness. MND New Zealand needs that help so that it can continue its support for those living with MND.

Whatever your connection to MND New Zealand, I thank you for the support you give.

Mā te wā,

Beth Watson



'Digital Legacy' Project

OLIVER CLABBURN's 'Digital Legacy' project focuses on people with MND recording short videos to be given to a child or young person in their family for use in the future. Over the past few months, Oliver has spoken to a number of people who have recorded a video legacy in various forms, such as personal childhood memories, messages of guidance, or, the telling of a children's story.

He is interested in the experience of creating a video legacy and looking to hear from people who are willing to take part in an interview with him over Skype.

He is also looking to speak to bereaved young people between the ages of 11 and 24 about their experiences of using videos after someone in their family has passed away from MND. These videos could be things like home movies of special family occasions, or videos that were purposefully recorded for the young person.

"My project is moving into its final year," says Oliver. "While I have spoken to a number of people, I would really like to hear from more. If you are living with MND and have recorded or are interested in recording a video legacy for a child in your family and willing to take part in an interview, please get in touch."

Alternatively, if you are a bereaved young person aged between 11 and 24 and sometimes use videos for support, Oliver would also love to hear from you.

For more information, email
clabburo@edgehill.ac.uk

Greetings from Grant



IN EARLY DECEMBER

I attended the annual meeting of the International Alliance of ALS/MND Associations in Dublin, Ireland.

The meeting was attended by 110 delegates from over 35 countries. It presented an opportunity to hear from groups in Russia, where attempts are being made to establish an MND association in Moscow, newly established MND associations in parts of South America and Asia, and mature organisations like ours with over 30 years' experience.

The meetings consisted of the Annual General Meeting, which covered two days, followed by a one day Allied Professional Forum. In addition there was a three day International Symposium on ALS/MND for health professionals and researchers. Given the specific content of the Symposium I did not attend.

Over the course of the week there were

a total of 1100 attendees. I was able to make contact with a number of people and discuss their work, and our work here in New Zealand. In general it appears that we are in a good position with regards to our service delivery model, that is the way we support people living with MND their carers and families, and the health professionals who support them.

It reinforced to me that our health system is one of the best in the world, with leading clinicians and health professionals across all disciplines providing high quality health care while not requiring individuals to fund their own care costs. Many of the attendees expressed their admiration for our health system and contrasted it very favourably with their own.

The Allied Professional Forum consisted of presentations around new technologies and specific areas covering care and other health related matters.

One of the most moving presentations was from Gudjon Sigurdsson, President of MND Iceland. Gudjon is living with MND and his presentation was titled 'Life is Now'. Gudjon impressed upon

us that while research is important, so is the support of people with MND and we must not forget them in the quest for a final cure.

This annual event is an excellent way for us to connect with the global MND community and both offer our insights to them and take away from others' learnings for our mutual benefit.

If you wish to view the presentations from the Annual General Meeting and the Allied Professional Forum please go to www.alsmndalliance.org/alsmndmeetings/past-meetings/2016-meetings/

Annual survey results

ANOTHER ANNUAL MND NZ Client Survey was conducted at the end of last year. We also conducted our first ever survey of health professionals. We were delighted that so many busy clinicians took the time to respond, and that 94% rated our Support Service as valuable.

Our clients told us that our most valuable services are our Support Service, newsletters, MND info pack, liaison with other service providers, and support for carers and family.

We used Survey Monkey to collect responses online for the first time. Using Survey Monkey saved us a considerable amount of time in collating responses, but we're aware that many of our clients are still more comfortable responding on paper, so we will continue to provide paper surveys in future to those clients who prefer not to respond online.

Both the healthcare professionals and clients who responded to our surveys are aware of the many demands on Support Worker time, and the large geographical areas their roles cover. As always, we would love to provide as much support as possible no matter where clients live, but our funding restrictions mean those in outlying areas are only visited in person occasionally.

We are investigating other ways to stay in touch. If you are interested in using Skype to connect with your Support Worker, please let them know and we can give it a go!



Gudjon Sigurdsson

Call for Symposium 2018 papers

THE 28TH INTERNATIONAL SYMPOSIUM ON ALS/MND will

be held in Boston, USA on 8-10

December 2017. The Symposium is currently calling for papers to be submitted for possible presentation at the event. The deadline is 26 May 2017. See www.mndassociation.org/research/international-symposium for more information.



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OF ALS/MND ASSOCIATIONS



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Please contact us:

- If you have any suggestions for inclusion in the next newsletter
- If your address has changed
- If you know someone who would like to receive our newsletter
- If you no longer wish to receive MND News or would prefer to receive by email.

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MND New Zealand Funders

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