# InTouch

KIA NOHO TATA Inclusive | Inspiring | Informative Summer 18 Issue 101


## Hello Summer
Enjoying the warmer months

## Get creative
The power of words

## Freedom News
Our annual campaign

## 'I choose' explained
Understanding the new career support

Muscular Dystrophy New Zealand

# InTouch

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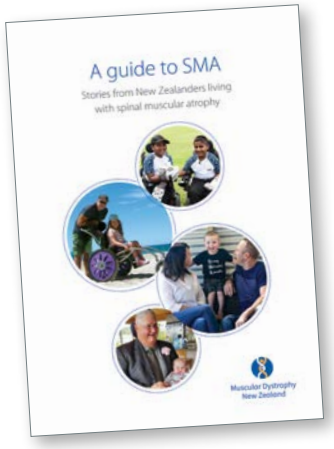
# Contents

## FEATURES










Muscular Dystrophy New Zealand

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## REGULAR



## MDA NEWS



## FREEDOM NEWS

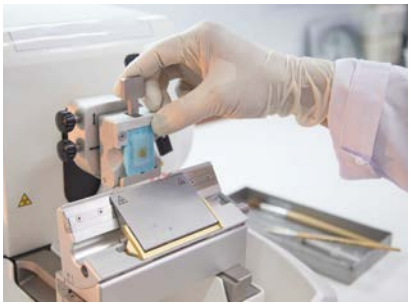


## RESEARCH





## YOUR CONDITION IN REVIEW





## ASK THE PANEL

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Also thanks to Allied Medical, Biogen and Sanofi Genzyme, the ARA Lodge No 348 IC Charitable Trust, the Clyde Graham Trust, NZ Post Community Post, Auckland Council, Richdale Charitable Trust and the Independent Living Service for their continuing support.

# Catching up with Trevor

Kia ora everyone, greetings to you all.

It has been a very busy time for the organisation and the National Council.

Thanks to all of the members and supporters who got behind the Freedom campaign this September. It has been a fun time, with fundraising and awareness raising happening across the country. You can read more about these events on page 8.

I am also pleased to inform you that MDANZ has officially become a recognised Disabled Persons Organisation (DPO). We were welcomed as the 7th member of the DPO Coalition in September and as part of this group, we now take a seat at the table with government in the disability-led monitoring of the New Zealand Disability Strategy, Disability Action Plan and the implementation of the Convention on the Rights of Persons with Disabilities (CRPD) in New Zealand. Increasing our member engagement will be a continued focus, so that we can ensure we are authentically representing our members in this forum, which influences government decisions.

As MDANZ approaches its 60th anniversary, the National Council have recognised that we need to review our organisational structure to help us achieve sustainability for the future and to ensure we are in the best possible position to continue to provide the services our members require. We look forward to engaging you in this process when the time comes to consult with you about potential options for a future organisational structure.

On behalf of the MDANZ, I also wish to farewell Ronelle from her role as Chief Executive. Ronelle has worked hard to manage the organisation and its funds efficiently, and established the Member's Discretionary Fund and Duke of Edinburgh programme during her three years in the role. We can wish her well for the future and thank her for her contribution. This role won't be replaced immediately, in light of the broader organisational review that we plan to carry out. However services will continue as usual to our members.

As we approach the Christmas break, I encourage as many members to get along to camps, Xmas get togethers or coffee groups being held in your region. I am looking forward to the Northern Branch camp and Xmas party, which every year, just seems to get bigger and better. At this time I would like to thank all of the staff for their hard work throughout the year and also, the members of the organisation's various national and regional committees and the research trust. We appreciate these members and supporters who volunteer their time to carry out governing roles for our organisation. May you all enjoy a nice break over summer.

Merry Christmas everyone,

Trevor Jenkin
*National Council Chairperson*

# In touch with Ronelle

Ngā mihi nui ki a koutou, warm greetings to you all.

I have spoken before about my grandfather George Howe, who was of Tainui descent and the eldest of nine siblings – the only one to have been born with FSHD. Being the eldest son, he was a leader and a pillar of strength for his family and community. Before he passed away in his early 40s from respiratory complications, my grandfather became a founding member of the support group, that later became the Muscular Dystrophy Association of New Zealand and registered as an incorporated society in 1959.

This organisation has been a part of my family's life for many years, and I have been privileged to have led the organisation as Chief Executive since August 2015. It has been an immense responsibility, because I stand on the shoulders of the leaders who have gone before me. As I depart from the role, I have two key thoughts in mind. First, I hope I have done you all proud and made a positive difference. The second is my hope for the future – which is that MDANZ stays true to its vision and organisational mission, that our leaders and staff take care of each other, and that we never lose sight of our purpose.

There is a Māori proverb or whakataukī that highlights the importance of humility.

Kāore te kumara e kōrero mō tōna ake reka.

*The kumara (sweet potato) does not say how sweet he is.*

However it is impossible to not celebrate the achievements of the past three years without being proud and shining a light on the collective efforts that have facilitated the development of our organisation and services, for the benefit of our members. Many gifted individuals have contributed to these achievements.

Ronelle's mother and grandfather.

I thank you all, with deep sincerity and wish you well for the future.

Kia noho tata, keep in touch.

Ronelle

Ronelle Baker
*Chief Executive*

# Haere rā Ronelle

After three busy, eventful and memorable years sitting at the Chief Executive's desk, Ronelle Baker is moving on. MDANZ staff and members want to say a huge thank you for all her hard work and significant difference she has made for so many people during this time. Here are just a few highlights of the past three years. Ronelle, you will be missed!



Clockwise from top left: Preparing to leap from the Sky Tower to launch our first ever Freedom Campaign; at Parliament with the Carers' Alliance; at a fundraising high tea with Minister Carmel Sepuloni and Sophie Tauwehe Tamati; with Disability Rights Commissioner Paula Tesoriero; with patron July Baily at the 2017 Freedom Cocktail Party; with rangatahi Camille Petersen and Olivia Shivas; doing Round the Bays; celebrating Good in the Hood results with the team.

# A cup of tea and a catch up with … Dympna Mulroy

Each issue we introduce a MDANZ team member:

**How long have you worked for the Muscular Dystrophy Association and what do you do?**

I have worked with MDA for six years. I started as the sole fieldworker for the Wellington Branch, covering the Lower North Island in 2012. I went part-time in this role in 2016 to undertake my Masters degree, where I am looking at Myotonic Dystrophy and employment. In 2017 I became practice leader for the fieldworker service in addition to part-time fieldworker. I most recently became the Member Services Manager at National Office in October. I took over this role from Miriam Hanna as a 9 month secondment. I am still trying to find my feet, mainly getting used to Auckland transport, but enjoying the change and new learning.

**What qualifies as a great day at work for you?**

The conversations I have with members. A range of emotions are exchanged from sad to happy to excitement to frustration or anxiety. People share a part of themselves with me and I feel honoured to hear their stories. There is no greater feeling then knowing you have made a difference and hearing the words "thank you", giving them reassurance or putting a smile on a person's face.

**If resources and funds weren't an issue, what would you like to see our members enjoying?**

Freedom to go where they want to go without having to worry about transportation costs, accessibility, social exclusion and toileting facilities. These are things many people without a disability take for granted.

**What's the perfect morning tea for an office shout?**

Freshly baked sultana scones still hot from the oven with butter and jam.

**What are you passionate about?**

I am passionate about equal rights for everyone. Everyone should be treated fairly with respect and dignity. I believe people should have the freedom to make their own choices, perform activities that bring meaning to their lives and be the best version of themselves. N

## Christmas Hours

National Office and the branches will close for the holidays on **Friday December 21st** and reopen on **Monday January 7th**. Check mda.org.nz for emergency contacts during this time. N

# Dukies update

It's been a while since we last updated you all on how our five superstars are getting on.

Our dukies will come together for the first time in Auckland from 30th November – 2nd December to take part in the Adventurous Journey component of the Duke of Edinburgh's Hillary Award. A variety of activites are planned that will challenge them to step outside their comfort zone, work together as a team, and try new things.

They've all been making great progress. Dylan has completed his VEX IQ Robotics course and is also continuing his service with dog petting at rest homes.

Ella has been volunteering at Riding for the Disabled (NZRDA) and perfecting her badminton skills. Instead of hairdressing, she has taken up sewing.

Ciaran has been recovering from surgery and is starting to move back into his routine. He is very keen to continue his Bronze Award and will be walking dogs, playing table tennis and trying air-rifle shooting.

Grace has been working hard creating puzzles and has finished cake decorating. She is now working on her gym routine and making sure she can tick off the goals she's set for herself.

Jack is our very first Dukie to finish off his chosen service, physical activity and a skill. Jack trained for six months to compete in the New York Marathon. He has also learnt to cook and hosted a radio show. N

Amelia Noyes

# DPO Update

MDANZ was formally welcomed at the September 2018 meeting of the DPO Coalition in Wellington.

The event was the culmination of a three month application and due diligence process, with Tristram Ingham and Ronelle Baker in attendance as the appointed board and management representatives for MDANZ.

Becoming a recognised Disabled Person's Organisation (DPO) in Aotearoa New Zealand is a significant achievement for our Association. Other DPOs currently recognised as part of the Coalition are the Disabled Persons Assembly NZ (DPA), Blind Citizens NZ, People First NZ, Deaf Aotearoa, Kāpō Māori Aotearoa, and Balance Aotearoa. We have joined these groups as the 7th member of the DPO Coalition.

DPOs have a mandated role to work with government to monitor the implementation of the Convention on the Rights of Persons with Disabilities (CRPD) in New Zealand. The CRPD is an international human rights treaty of the United Nations, established to protect the rights and dignity of persons with disabilities.

If you have heard of the slogan "Nothing about us without us!", you may recognise this has been used in the context of social change, as disabled people seek greater influence over policy and decisions that impact their lives.

The New Zealand government has an ambitious agenda, and government agencies are seeking increased participation from the disability community to both shape the political agenda and to deliver prioritised objectives under the CRPD. They need a strong representative channel to converse on significant issues and strategies, and that's where the DPO Coalition comes in.

As a member of the Coalition, MDANZ will now play a role in the disability led monitoring of the CRPD, co-governance of the Disability Action Plan, and contribute to discussion about many other policy areas such as statistics, housing, education, justice and transport. We look forward to representing your views at the highest levels of government and will have the opportunity to meet with the Office for Disability Issues, government officials and Ministers, to raise collective views around important issues.

This is an incredible opportunity, and to be effective in this role, we need to make sure that we stay connected with you, our members, and ensure that you have access to information about the work of the government and the DPO Coalition. We've started a new webpage so that and you can view updates online, and click through to various surveys that may be available. This is located under the member's area tab of our website.

We will also keep you informed through updates in In Touch magazine, and you may find that we email you a bit more often, because we want to offer you more opportunities to have your say and participate.

As former Green MP Mojo Mathers said when she spoke at our 2016 AGM, "The CRPD is important to everyone. When everyone participates, everyone benefits." It is our time to speak up, get involved, and make a positive difference in this space.

Find out more at www.odi.govt.nz or www.mda.org.nz/membersarea/DPOCoalition

Tristram Ingham and Ronelle Baker (front; left and centre) are welcomed by DPO Coalition members.

# The diagnosis experience

Understanding what it's like to be diagnosed with a genetic disorder.

Little is known about what people experience when being diagnosed with a genetic disorder and which factors may influence their experience. In order to inform policy on the delivery of genetic diagnoses and improve the experience for patients with rare genetic disorders, people with genetic muscle disorders were asked to describe their experience in a recent study.

This study, Patients' Experience of Diagnosis of a Genetic Muscle Disorder is a sub-study of MD-Prev and was contributed to by MDA's research arm.

A question designed to understand the diagnosis experience asked firstly, whether the participant had a genetic diagnosis, what their experience was in relation to the testing prior to receiving their diagnosis, their experience during the diagnosis, what their experience of any follow up that occurred, and their overall experience. Responses were analysed using a mixed methods approach.

Questionnaires were delivered to 803 adults, either in person by face-to-face interview or by phone, or online, or by paper form. 501 responses were received. (62% response rate).

Prior to being tested themselves 41 percent knew of family members with symptoms of the disorder and 31percent knew at least one family member who had tested positive for the disorder. Despite being familiar with the disorder, these respondents also felt they received too little information at the time of diagnosis and did not receive sufficient follow-up after receiving the diagnosis.

Regardless of prior experience with the disorder, the diagnosis of a rare genetic muscle disorder is a traumatic and difficult experience for most people. Support from family was identified as the most helpful thing during and following diagnosis. All participants felt they were provided with too little information at the time of their diagnosis and many did not receive sufficient follow up. N

# Successful funding applications

Grant recipients for the 2018 grant funding round of Neuromuscular Research New Zealand are;

- Kathryn Stowell for work on functional characterisation of ryanodine receptor variants associated with malignant hyperthermia
- Fiona Graham for a mixed methods study into tele-health wheelchair and seating assessments
- Sophie Burling for innervated muscle-on-a-chip for improved diagnostics and investigation of malignant hyperthermia
- The NZ NMD Registry. N

Freedom

Our month of Freedom was a great success. Thank you so much to everyone who helped out all over the country. With your help, we raised much needed funds for MDA and our members, and raised awareness in the community about the experiences of our members and the conditions we support. Here is a brief round up of what went on around the regions.

## Northern

Northern Branch kicked off the month with a sponsored morning tea for MD, wearing red to acknowledge DMD Awareness Day on 7th September. Members of the Northern Branch held a stall at the Matakana Markets and were supported by the local community who donated baked goods for sale. A street collection was held in Auckland's CBD, and at the end of the month, on LGMD Awareness Day, a large community of supporters dressed in lime green, walked around the Hatea loop in Whangarei, to raise awareness and close the month long campaign.

support@mdn.org.nz

From top: Street collection; Matakana Market; morning tea in red; walking the Hatea loop.

Top: Minister Carmel Sepuloni makes a donation at Wellington Airport. Above: At the blossom parade.

## Central

The campaign was a phenomenal success for the newly-renamed branch, with more funds raised than ever before. Volunteers were on hand to help out at street collections and special events such as the Hastings Blossom Parade. It was a slightly larger group than last year due to the willingness of a few volunteers from Karamu High School, so a big thanks goes out to Nathan and Macayla Parkins and friends for joining in and holding up the banner, made by Jarrod, who also gets a big thank you.

The name change to MDA Tuaatara | Central Region, or Central Region for short, was confirmed at a Special General Meeting. The renaming demonstrates branch commitment to better representing the entire Lower North Island region and upholding the values of the Treaty of Waitangi. The Te Reo Māori name Tuaatara refers to the spine of the fish – Maui's fish – the mythological name for the North Island.

office.mdawgtn@xtra.co.nz

Top: Ronelle Baker with Prudence Walker. Above: Ronelle with Dr John Alchin and Simon Goss.

## Christchurch

We had two major areas of focus in Christchurch for fundraising – a sausage sizzle at Bunnings and shopping mall collection which was supported by students from Christ's College. Thank you for giving up your time to support MDA Canterbury this September!

The Commodore Airport Hotel supported our third, annual education seminar held in collaboration with National Office. Members came from as far as Franz Josef Glacier to attend the session. Simon Goss and Dr John Alchin from the Pain Management Centre at Burwood Hospital gave an informative talk on managing pain, and Prudence Walker from CCS Disability Action shared her story and spoke about ways to get involved in disability leadership.

mdacanty@xtra.co.nz

## Southern

This year's Freedom campaign was all about our young community members and having fun. Special thanks to the local schools who supported the campaign with activities and mufti days. Our newly formed Lego Club continues to be a huge hit with budding builders in the South and we hope to grow this in 2019.

joanne@mda.org.nz

Having fun at school and playing with lego.

# Hello Summer!

## *Bring on the warmer weather and longer days*

There's so much about this time of year that simply makes life easier. We speak to members to find out what they love about summer.

### Serena and Peter Leong-Teo

Summer is always a favourite time for Serena Leong-Teo who particularly looks forward to the longer days. But this year is going to be extra-special for Serena and her husband Peter, because they are going on a "second honeymoon" to celebrate 30 years of marriage.

"We were going to retrace our first honeymoon in New Zealand, but since I've been using a wheelchair, it is easier to go on a cruise. So we will be cruising New Zealand, mainly the eastern ports, with Royal Caribbean for 10 days, ending up in Sydney. We chose this itinerary because they go around the Sounds and they had a mobility room available."

Alongside the cruise, they are planning a family holiday to Bayleys Beach and are looking forward to exploring a place they haven't been before.

Christmas is another highlight of summer, and Serena says this time of year is about the birth of Christ, rather than Santa for her family.

"We celebrate Christmas by having a festive reunion dinner on Christmas Eve. Then we go to church. At midnight, if we're still awake, we open Christmas presents. Nowadays, we tend to go to sleep first and open presents later. We're growing old!"

MD Northern Branch camps are always regular events on the couple's calendar and they enjoy the chance to catch up and connect with other members.


Serena and Peter are looking forward to celebrating their 30th wedding anniversary this summer.


It will be all about table tennis for Ashvin and Arvinth this summer.

Serena won't be drawn on her favourite summer ever, she likes to look forward, rather than back.

"The best is yet to be," she says. "The summers we remember the most are the ones where extended family comes and we tour around New Zealand."

*MD Northern Branch camps are always regular events on the couple's calendar and they enjoy the chance to catch up and connect with other members.*

## Ashvin and Arvinth Sathiyaseelan

Christmas is coming early for Ashvin and Arvinth Sathiyaseelan. The 12-year-old twins are sports mad and while they have a love of cricket, they can no longer participate fully. These days it is boccia and table tennis that has given them a competitive focus, and they are beginning to attract the attention of para-sports coaches as they out-play more experienced players in regional and national championships.

This summer, Ashvin and Arvinth will be working with the New Zealand Para table tennis coach, attending a national training camp in November, and getting in loads of practice time at home.

A table tennis table has been ordered, and mum Dharshi explains that her biggest challenge is to find a space big enough to fit both the table and two power chair using players, before it is delivered to their home. The other challenge will be picking up all of the balls when they hit the floor!

The twins have a built in partner for practice sessions and enjoy competing against each other at times. The future looks bright for these young competitors who are staying active, having fun and achieving success doing something they love.

## We want to see your summer!

Summertime means different things to different people and we'd love to see what it means to you. Take a photo over the next few months and email to info@mda.org.nz and we'll put a collection together.

Lydia and her Mum at the FARA family forum in September.

# Keeping the faith

## *New Plymouth's Lydia Wilson has a very special outlook on life.*

Mother Teresa once said, "Every time you smile at someone, it is an action of love, a gift to that person, a beautiful thing."

Such a small gesture can have enormous effect, and when you meet Lydia Wilson, you cannot help but be uplifted by her smile and positive energy.

When asked about why her faith is so important to her, Lydia will tell you that it is impossible to consistently have a genuine, joyful and positive attitude if you don't know the God who made you.

Faith has given Lydia an inner strength, which helped her strive toward self-determination, following a difficult time as a young adult. After an acute episode that resulted in hospitalisation, Lydia's needs were considered too high to be met at home with her parents on the family farm. She was struggling mentally and physically, and with few options available to her, Lydia found herself in a rest home at the age of 19.

It wasn't an easy decision for her family to make and for Lydia, it meant starting on a parallel journey of rehabilitation and spiritual growth. Using her voice to advocate for herself, she has achieved a more independent life with more joy, meaningful connection, choice and control.

Having been a resident at Te Henui Lodge, that has six wheelchair-friendly rooms, for three years now, Lydia is proud to be able to live in her own place, manage her food and dietary needs, doing things at her own pace and engaging with Christian fellowship more than twice per week.

Social life is busy and revolves around her faith - whether it be a women's bible study group every Tuesday, or young person's group every second Friday, Lydia loves

to teach, learn and reinforce her Christian values through these interactions.

Faith is a verb not a noun for Lydia. It is something she lives, teaches and actively encourages to everyone she meets.

## 10 questions for Lydia

1. **My favourite way to spend a day is …** Waking up at 5am, having a shower independently, praying over Skype, and walking with carers using a walker, before going downtown on my Powerchair at 7.45am to speak to people about Christ and give out tracts while people are on their way to work or school. It is hard work, and even harder to decide to do. But this is a very rewarding use of time. You meet all kinds of people, and give them hope and purpose through the cross.

2. **The best advice I have ever been given is …** Have a break from work (by Mum).

3. **My favourite quote is …** "Commit your way to the Lord. Trust in Him and He will act." Proverbs 16.3.

4. **My all-time favourite movie is …** I don't watch many movies now, but I cried over Charlotte's Web at primary school. I was so emotionally involved. I remember Wilbur the pig.

5. **My most prized possession is …** Probably my cellphone. I can do so much with it.

6. **My favourite food is …** Wraps with hummus and kabuli and tomato and beetroot and ham.

7. **If I'm ever feeling down, the best way to cheer me up is …** Make me want to go and talk to someone about Jesus, and cheer them up if they are down. This forces me to get over my own problems and become brave. This boosts my emotions.

8. **The thing I love most about summer is …** Having warm legs all the time, without blankets.

9. **For me, Christmas is all about …** When we remember Jesus came down to earth from Heaven to save us from ultimate death.

Lydia loves getting out and about in her Cogy and is thankful for contributions from the Lions, Rotary, Red Cross and a Givealittle page that all helped pay for it.

*Faith is a verb not a noun for Lydia. It is something she lives, teaches and actively encourages to everyone she meets.*

10. **I'm glad to be part of MDANZ because …** We get wonderful people to work for us and provide funding grants and write articles, and we get optional catch-ups with a fieldworker.

# Making a difference

## *Sharing knowledge, tears, laughs and beers*

Dion and Trish Ahern, parents of Harper who has Duchenne muscular dystrophy, attended the 2018 Duchenne ACTT Now Conference in Sydney in September. They came back with plenty of new facts and information, and just as importantly, they made valuable new networks and friendships. Here are Dion's notes from two inspiring days.

I've been to a number of work-related conferences in the past, and had always come away with useful learnings and experiences, but we had never been to something quite like this. We weren't quite sure what to expect, and hoped it would be valuable for us.

It was also a good opportunity for Trish and I to get away together without the kids (which is important), and for the kids to stay with their Nan and Pop (which is important too!). We'd lived together on Sydney's Northern Beaches during our early 20's, so it was a nostalgic few days for us before the conference began.

### The conference

The two-day conference was run by the *Save Our Sons (SOS) Duchenne Foundation*, a non-profit organisation whose aim is to not only to find a cure for Duchenne, but to *"make a difference in the lives of all living with Duchenne across Australia"*.


We were nervous as we registered and collected our 'goodie bags' on the first morning, and were warmly welcomed by Carolyn Campbell-McLean. Carolyn, an inspiring and purposeful lady who lives with a disability herself, told us about her husband who had DMD and lived into his 40's. "Staying positive and active", was his secret she said. Hope, I translated.

The programme was diverse and comprehensive, with excellent speakers presenting topics about respiratory health, bone health, physiotherapy, dental care, cardiac care, steroid treatments, fracture management, genetics, neurobehavioral issues, wheelchair technology, mental health, clinical trials, etc etc.

It was also confronting, and by the end of Day 2, Trish and I were overwhelmed and knackered!

We had absorbed heaps of information and facts, which we are still processing.

Some of our key takeaways are;

### Clinical trials and CRISPR gene therapy

- There are many DMD clinical trials in progress and on the horizon; Australian clinics and researchers are assessing and deploying many of these
- Clinical trials require quite a commitment from families
- CRISPR has exciting potential for Harper and others with DMD!
- CRISPR is quick, accurate and affordable – yet trials on humans, and deployment widespread will still take years as the 'off-target' effects are still largely unknown (you can't reverse gene therapy)
- CRISPR is not just for particular genetic mutations (like

exon 51 skipping therapy) – this is good news for Harper

- CRISPR won't reverse the effects of DMD, but will stop muscle degeneration – so the earlier we and others can have this, the better
- We need to be aware of the clinical trials on the horizon, and get onboard as soon as possible

## Respiratory care and management

- Preventing and managing respiratory muscle weakness is absolutely critical to Harper's survival
- A simple cough for a person with DMD can quickly become a severe infection, as weakened respiratory muscles struggle to clear mucus
- Specialist cough assist machines and ventilation equipment will likely be needed when Harper is older – these technologies are improving, and becoming less invasive
- Respiratory is not just about being able to breathe oxygen in well, but also about breathing out and expelling carbon-dioxide from the body (which can cause issues if not expelled well enough)
- People with DMD need to avoid inhaled anesthesia and take extra care if supplemental oxygen is required for any reason
- We need to be performing annual Pulmonary (lung) Function Tests for Harper now, which includes spirometry tests, peak cough flow tests, sleep studies, etc.

## Behavioural and learning issues

- Roughly 40 percent of kids with DMD experience neurological learning difficulties caused by lack of dystrophin in the brain
- Learning (and mental health) issues common in DMD kids include anxiety, ASD, ADHD, depression, dyslexia, dyscalculia, short-term memory and attention
- School is super important for kids with DMD to help develop their mental health, resilience and social lives, as physical activities become limited and skills such as reading, writing and IT occupy their lives
- We need to continue ensuring Harper doesn't fall behind at school, and act quickly if we are seeing learning / behavioural issues

The conference provided valuable networking opportunities.

*CRISPR won't reverse the effects of DMD, but will stop muscle degeneration – so the earlier we and others can have this, the better.*

## Steroids, bones and boners! (puberty)

- The benefits of steroid treatment for DMD are "undisputed", and outweighed the side effects (e.g. poor bone density, weight gain, delayed puberty, restricting how tall a kid grows, etc)
- DMD kids using Deflazacort on average, lose the ability to walk at around 14 years old, whilst kids using Prednisolone stop walking around 11 years old (this is a significant difference which we weren't aware of)
- DMD kids on steroids are three times more likely to break their bones than other kids, and will often have unusual and severe fractures
- 50 percent of DMD kids will break a lower limb, and 50 percent of these kids will never walk again
- Aggressive management of leg fractures is needed; i.e. surgery as opposed to being immobile in a cast for too long

- The most common cause of fractures for DMD kids is falling out of their wheelchairs from not wearing seat-belts (this has happened to Harper once already!)
- Osteoporosis and vertebrae fractures are common – regular spinal x-rays / imaging are needed
- Steroid treatment suppresses the adrenal gland – which normally triggers puberty
- Puberty is not only important for social wellbeing and self-esteem, but also for healthy bones (puberty increases bone mass by 50 percent)
- Delayed puberty is universal for kids with DMD, so testosterone medication for boys should be considered from the age of 13 to help kick-start puberty
- Steroids impact how tall a kid can grow, but there is opinion that being shorter is actually beneficial for DMD people's muscles and limbs (growth hormones aren't commonly used)
- We need to investigate further the use of Deflazacort instead of prednisolone (Deflazacort is not funded in NZ), develop an Emergency Management Plan for Harper, ensure specialists are engaged in the aggressive treatment of any broken bones and perform 1-2 yearly DEXA tests (measuring bone density) alongside spine imaging/x-rays

## Our wellbeing

- For families dealing with DMD, there are high rates of depression/anxiety
- Friends don't always know how to help; some drift off, and some new friendships can develop
- It's important to actively seek friendships and maintain social relationships with others
- Caregivers need to make sure they make time for themselves, and exercise!
- Don't let DMD define our lives!
- We need to set some family goals, acknowledge and allow for each other's (and our families) grieving styles, beware not to add additional responsibilities to Mia, and make each other a priority

## Other titbits

- Nutrition for Harper is important, and it may be worth engaging a dietician in the near future
- Kids on steroids only need to eat about 80 percent of kids their age; especially if they are in a wheelchair full-time and less active
- Cardiomyopathy (heart disease) occurs universally in patients with DMD
- We need to continue with regular ECG's, and explore further performing cardiac MRI's and other preventative treatments such as ACE-inhibitors and beta-blockers, as well as encourage Harper to perform stretches more often at home!

## Networking value

We met some awesome people! Whilst there were facts and information galore that has improved our knowledge immensely, and empowered us slightly… perhaps the best thing for us the networking experience with others.

We met parents in similar situations and stages to ours. Parents who had only just received the diagnosis of a child. Parents with two or three kids diagnosed. Parents whose children had moved away as independent and successful young adults. Parents who had moved away from each other due to the stresses involved and parents whose children had passed away far too early.

We met driven, inspiring and purposeful people. Industry professionals passionate about their chosen field, and who really wanted to make a difference. Sponsors who weren't just on another product sales trip…

We shared knowledge, tears, laughs and beers. And from the moment Carolyn introduced herself to us on the first day, through to the moment we scampered off to our airport shuttle, compassion, love and understanding was abundant.

So yep, this trip was valuable for us. More so than we expected. Thank you to all of those involved who helped make it happen, the sponsors who supported this, and to families and new friends we've met and made… see y'all next time.

Dion and Trish Ahern.

# The power of words

## *Why writing helps in so many ways*

Northern Branch Fieldworker Darian Smith is a published author who mainly writes fantasy. He shares advice for new writers.

Writing is a hugely beneficial and often underrated art form. People have been using writing to process their thoughts, share their experiences, and tell stories for as long as there has been literacy - perhaps even before, if you count cave paintings. A picture might be worth a thousand words but words have a specificity and power that pictures do not. They help us understand ourselves and the world around us.

Writing is also a creative outlet that requires very little in the way of physical ability. Jean-Dominique Bauby famously wrote The Butterfly and the Diving Bell by blinking his left eyelid. With today's technology, it's much easier.

Writing can be therapeutic too. Keeping a journal, writing your life story, or even creating a fictionalised version of your experiences can help to process how you're feeling in a safe way.

Like any activity, there are skills to learn but anyone with a basic grasp of language can write and gain the benefits of doing so. A good place to start is to pick the type of writing that suits you.

### Blogs and Articles

These are non-fiction pieces of writing intended to educate and share information. They enable the writer to talk about a specific topic or experience, to organize their thoughts, and to share what they have learned, thus making a difference in the lives of others. It's a good idea to keep to a single topic and keep the writing short and easy to read. The more simply you can convey an idea, the easier it is for readers to understand it. You may want to seek a magazine or website to publish the article, or set up a blog yourself.

### Journaling

The main purpose of keeping a journal is to process thoughts, feelings, and events. Journaling can be very therapeutic. A journal doesn't ever need to be seen by anyone so it is a very freeing form of writing. You can say whatever you think or feel without fear of repercussions or causing offence. One way to do this is to clear your mind and write whatever comes into your head with no judgement. Journaling is most beneficial when focused on thoughts and emotions, rather than just a diary of the day's events.

### Fiction

Fiction can be used to explore, entertain, and contribute to social awareness. Fiction could include novels, short stories, movie scripts, or plays. With fiction you can take things to extremes and explore them in ways you would never do in real life. It can be an escape into another life with wonderful new experiences, or an exploration of a dark emotion in yourself from the safe distance of being in a fictional character's world. Fiction can help show the world what it is like to live a particular life and change readers' attitudes as they walk a mile in the character's shoes.

### Autobiography

An autobiography tells your life story and shares your experiences – ideally both the internal experiences and the external ones. Combining the benefits of a blog and a journal, this is often a popular choice for writers, particularly later in life. Unless you're already famous, however, it is likely to have a smaller, niche audience.

Whatever style of writing you choose, the key is to keep at it. Don't let thoughts of your intended audience hinder you in your first draft. Just write and see what happens. The wonderful thing about writing is that you can always change it later. The most important thing about writing is to enjoy it!

# Telling our stories

We were delighted to launch our new SMA resource at an SMA family day held on at our National Office in September. A big thank you goes to the families who shared their stories in the booklet.

A guide to SMA will help raise awareness of spinal muscular atrophy (SMA) and assist with lobbying for access to new breakthrough treatment Spinraza (Nusinersen). Biogen, the company marketing Spinraza, has achieved Medsafe approval and has now applied to PHARMAC for funding consideration. This application is now visible on PHARMACs website www.pharmac.govt.nz/wwwtrs/ApplicationTracker.php?ProposalId=1694 and we have been informed by Biogen that the application will be reviewed by the newly formed Rare Disorders Subcommittee and Pharmaceutical Therapeutic Advisory Committee (PTAC) before funding decions are made. We will keep you informed as new information comes to hand about this process.

The Expanded Access Programme (EAP) which Biogen is funding, and working in partnership with Starship Children's Hospital to deliver, will provide compassionate access to the most at risk children with SMA Type 1. We are heartened to know that our most vulnerable children will soon be receiving treatment under the EAP.

Overall, this is a very exciting time in the neuromuscular field, as Spinraza is proving to be a game-changer internationally. We are committed in our resolve to making this treatment available to all New Zealanders living with SMA.


A PDF version of this book is available on our website www.mda.org.nz. Here is an edited version of the story Colin Cockburn shares in the booklet.

## Overcoming the odds

Colin Cockburn has always been a movie buff. He has spent many hours watching and enjoying movies in his local theatre and seeing how the audience reacted to the protagonists as they overcome impossible odds. He made good use of this expertise as the manager of a video rental store for some years, gauging which movies would be a hit when they finally made it to tape. Colin himself, has overcome what many would have deemed, more difficult odds than most movie heroes would ever face.

Back in the 1960's, it was noticed that 5-year-old Colin wasn't running like the other children at school. The doctors investigated and believed he had Duchenne muscular dystrophy. His parents were told he would be dead by age 20 and were devastated by the news.

But that didn't stop Colin. While physical activity became more and more difficult, upon leaving school he started his own signwriting business. He began using a wheelchair at age 18 (late for someone with Duchenne in those days) but kept working. Recalling that time, he says, "Around age 25 I thought, I don't feel like I'm going to die tomorrow so maybe the diagnosis is wrong." He was seen by another specialist who rediagnosed him with SMA type 3 and was told to expect a much longer lifespan.

"Suddenly there was a future to plan for after all," he says. He'd never heard of SMA before but he connected with the Muscular Dystrophy Association for support and information. Colin set about making the most of the unexpected lifespan.

Now in his 60's, Colin is retired. He has carers to help him get up, shower, and prepare his food as he can't do these physical activities himself. His mind, however, is as sharp as always and his days are filled with social contribution, as he is on a wide range of community committees, disability

focus groups, a men's social group, Probus, and more. He's an active member of his church, playing a role in services and managing building maintenance. He regularly enters submissions into the Council's Long Term Plans.

Clearly, Colin loves being involved in his community, and provides more than many non-disabled people would, yet he has found it difficult at times. He recalls having his carer hours cut in the 90s and having to deal with 24 pages of questions every year to maintain the level of assistance he needed, despite the clear knowledge that his condition was progressive and not improving. He's noticed the system change for the positive in recent years.

"Since the UN passed the bill of Disability Rights, people have become more aware and it made me more aware. People's attitudes have slowly improved." He says working with the Council changed at that point too. An accessibility committee was established and he's proud of the work they've been able to do, getting an audit of the business district and bringing about changes. Colin thinks a big part of what's needed to create change, is for people to overcome the interpersonal barriers between the able bodied, and those with disabilities. "You have to get to know people and let them get to know you. People are a marvellous asset. Mix with them. It's important for them to see you as a person and know what you're all about."

Attitudes toward disability are not the only thing Colin has seen change over the years. "Technology has changed over time wonderfully," he says. Colin's first electric wheelchair was essentially a manual chair with a motor on each wheel powered by a car battery. He says it had a high risk of flipping over when in use. These days, he has one that is reliable, stable, with added features like leg raising and recline, and a clear indication of how much battery power is left. He says the innovations have been brilliant. Even using a dictation program for his computer rather than having to type is an advancement that has made life much easier for Colin.

One frustration that lingers around the technological advancements is the disparity in funding for it. Colin is frustrated that people whose disability comes from a genetic or medical condition are so much less able to access funding and support than those who gain their disability through an accident and are taken care of by ACC. "Treat people with long term chronic conditions exactly the same as ACC people," is his message for government. "That's the benchmark you should aim for. So there's access to everything they need."

Colin Cockburn and his sister, Jenny McDonald, enjoying a trip on Lake Taupo.

*"Suddenly there was a future to plan for after all".*

Access to treatment is another important message, and Colin describes the advances of recent new drug therapy as; "Marvellous for young people. I've never been one for getting into miracle cures but this seems to be the most positive new drug therapy and scientific treatment advance in my lifetime." He thinks perhaps the developments might be too late to make a big difference for him, but if given the chance he would try it. "If I could stop the decline of my fingers, that would be marvellous."

After a long life with SMA, Colin has a strong message for those just coming into the journey. "Don't live your life for your disability. Live it for your life. Get schooling, get equipped. "

There's a new movie theatre being built in Taupo and Colin's input is helping to make sure it's accessible. You'll see him there as regular as always, doing what he loves. Because no matter what difficulties might get in your way, he says, "There's always a solution."

# Living and working with myotonic dystrophy

Dympna Mulroy shares an update on her research project.

## Background

Some of you will know that I am completing a Masters Degree on the factors that influence work performance for people with myotonic dystrophy (DM). I am half way through my second year and am planning on submitting my thesis in March 2019.

I recently had the privilege of presenting at the Occupational Therapy Clinical Workshops in Napier in September. I am a trained Occupational Therapist and thus concerned with functional ability and purposeful activity. Work is one of the fundamental occupations that help define who we are in society It creates a sense of identity and purpose, and gives Occupational Therapists an important place in vocational rehabilitation practice. This is why I chose employment as my research focus.

I was somewhat surprised at the amount of attendees at the conference that didn't know about myotonic dystrophy. Myotonic dystrophy is one of the most common muscular dystrophies, albeit there was still only 343 people identified with the condition in the MD prev study.

Myotonic dystrophy (DM) is an autosomal dominant disease caused by an expansion of a DNA triplicate known as CTG. Generally, the more repetitions, the more severe the condition. Myotonic dystrophy is characterised by progressive muscle wasting and weakness.

Dympna receives a koha after her presentation.

Many of you participated in the MD Prev Study that ran over 2015 – 2016. A lot of data was gathered through the study questionnaire but all of it was not explored in depth including the section on employment. I was interested in what factors influenced work performance for people with myotonic dystrophy taking into consideration current literature pertinent to DM. I therefore extracted the demographic, diagnostic information and questionnaire data for adults (aged ≥16 years) who had been diagnosed with myotonic dystrophy type 1 and 2 for the purposes of my analysis.

The primary aims of my study were to:

1. Explore the rates of employment (defined as the number of people in paid employment) in adults diagnosed with myotonic dystrophy.
2. Determine levels of work performance in adults diagnosed with myotonic and time loss.
3. Identify the factors that influence work performance in adults with myotonic dystrophy.

## What we already know

The current literature identified that the employment rate differed across the type of conditions. Some studies showed that people with DM had lower employment rates in comparison to FSHD, SMA, HMSN, and LGMD. However there was no known figures of employment for

people with NMD in New Zealand or Australia. Although many studies investigated the employment rates of people with muscular dystrophy there was limited focus on factors affecting work performance and work loss. I am finalising the analysis but this is an overview of my findings to date.

## Results

This was the first known study examining what factors impact on work productive for people with DM in New Zealand. This study identified that there was a low employment rate (34.2 per cent) amongst this population which was lower than the national average of 58 per cent for people with a disability. The most common occupations were classified as professional, followed by clerical and administrative. The majority of the sample were of a working age with 57.4 per cent under the age of 50 years. Those that had worked, retired at a young age, and believed their condition affected their ability to seek or maintain employment.

A positive finding is the majority of those working are doing very well. Participants who sought adaptations stated that flexible working hours and reducing work hours were of most benefit. Others required physical equipment aids for work. Working with employees and employers that were understanding to the person's limitations and needs was also an important factor.

Others expressed low self-efficacy in getting work. They felt people would not want to employ them due to their condition and others stopped applying for jobs based on negative past experience. Many wanted to work but felt they were unemployable because they had DM. These environmental and attitudinal factors could be addressed with the right education and training.

An important factor shown between those working and not working was education. Those who had further education beyond secondary school were more likely to be employed. Due to the progressive nature of this condition and the occurrence of symptoms often at working age, support to pursue suitable jobs at a young age are important.

The mental/cognitive aspects were more of an impact on productive loss and performance than physical capabilities. This was likely due to the fact that people could adapt easier to physical difficulties than cognitive ones. Cognitive dysfunction can often determine levels of disability in activities of daily living more than physical.

The main factor affecting work performance was fatigue, followed by cognition, pain and mood. Fatigue impacts on all aspects of function and correlates with cognition, pain and mood. Thus focusing therapy in this area could be of major benefit for people with DM. Some of this input may need to be directed at employers to accommodate flexible working environments to manage some of the fatigue.

The unknown rate of progression can cause uncertainty for employers as well as employees. However it is encouraging to know that those in employment are doing well. An overall area of concern, is the low rates of employment for people with DM. Career guidance at an early stage could be good for this population group to help guide them into suitable roles and foster continued professional development. Knowing people who are working can perform well with appropriate support is hopefully an incentive to encourage more people into paid employment, and improve participation rates in the workforce which will have broader positive impact in people's lives.

# Treatment for Charcot Marie Tooth disease

Pharnext has announced Positive Results from Phase 3 Trial Using PXT3003 to Treat CMT1A.

Pharnext Pharmaceutical company announced positive topline results from its pivotal Phase 3 clinical trial (PLEO-CMT), evaluating two doses of PXT3003 compared to placebo during 15 months for the treatment of Charcot Marie Tooth disease type 1A (CMT1A).

PLEO-CMT was a pivotal, 15-month, double-blind Phase 3 study that assessed the efficacy and safety of PXT3003 compared to placebo, for the treatment of patients with mild to moderate CMT1A. The study evaluated two doses of PXT3003 in a randomised group of 323 participants age 16 to 65 years old. The primary endpoint was the Overall Neuropathy Limitation Scale (ONLS) measuring patient disability. Pharnext reports that a reduction of 0.3 point on this scale was determined to be meaningful according to previously described methodology.

Compared with placebo, a mean reduction of 0.4-point ONLS (95% CI [0.1,0.6], p=0.008) was observed in the trial group receiving the highest dose of PXT3003.

Additionally, PXT3003 was safe, well tolerated and showed a similar safety profile as seen in the Phase 2 study. In this study, PXT3003 provided first evidence of a meaningful improvement of CMT1A effects. Based on these results, Pharnext intends to file for market approval in the U.S. and Europe.

# Scientific publications vs lived experience

An interesting question asked at the recent World Muscle Society meeting was 'Do scientific publications fit with the expectations of individuals with myotonic dystrophy?'

A French group carried out a systematic review and comparative study seeking to answer this question and found that over the past few years the scientific knowledge on the myotonic dystrophy phenotype and disease progression has greatly improved. In parallel to this, patient reported data and rare diseases foundation-driven initiatives have contributed to better address patient needs and expectations.

They explored the relevance of current myotonic scientific research areas by designing a study that aimed to compare the research areas of interest reported in literature with; disease symptoms prevalence, and patients' expectations.

They found that scientific publications topics do not fully cover the range of symptoms most frequently experienced by people with myotonic dystrophy, either from clinician objective measures or patients self-reported measures.

In particular, researcher topics mainly focus on disabilities and life-threatening conditions such as heart-respiratory defects, muscular disability, and cognitive impairment. Conversely, other disease domains, including dysphagia, digestive tract dysfunction, fatigue and pain are under-represented in the scientific literature with regard to the frequency of patient complaints.

It is hoped that these results may support stakeholders and scientists to plan studies fitting more with the expectations of people with myotonic dystrophy.

# Can CRISPR treat DMD?

You hear a lot about 'CRISPR' in the news these days.

A recent Late at Auckland Museum event asked the question 'CRISPR – Utopian or Dystopian?'

A panel of four – two scientists, a bio-ethicist, and a professor of Māori and Indigenous Studies, discussed whether it would be a tragedy not to take advantage of CRISPR gene-editing and can CRISPR help all of us or will it be just the most privileged who might benefit.

The discussion ranged across multiple potential uses of CRISPR in the horticulture and agricultural industries, but it was in the area of human health where there was in fact the least controversy. The treatment of rare conditions, such as Duchenne muscular dystrophy (DMD), was provided as an example where treatment using CRISPR would likely be considered ethical and in fact unethical to not use it, if it were available.

But first, what is CRISPR?

CRISPR is a gene editing tool discovered only a few years ago and now being used in laboratories all over the world in plants, insects and animals. Progression towards using CRISPR to treat human diseases is moving fast.

It is an acronym standing for Clustered Regularly Interspaces Short Palindromic Repeat, which is a pattern of genetic sequences in DNA that can be recognised, cut, replaced or edited by a special cutting protein. The most commonly used cutting protein is called Cas9, hence CRISPR-Cas9.

Can CRISPR be used to treat DMD?

As published in Science in October, researchers used viral vectors to deliver CRISPR directly to the muscles of two dogs that were unable to make functional dystrophin. They found that after six weeks of treatment, the animals produced the protein at around 60 percent of normal levels in some muscle fibers, and microscopic examination showed that muscle integrity had improved.

When the team next administered the viral vectors into the bloodstream of another two dogs, the animal receiving the highest dose produced dystrophin at up to 70 percent of normal levels in skeletal muscle after eight weeks, and 92 percent in heart muscle. The work is still a long way from being applied to humans but is focused in that direction, with longer-term trials in animal models planned, and this offers hope for DMD.

# Inclusion body myositis

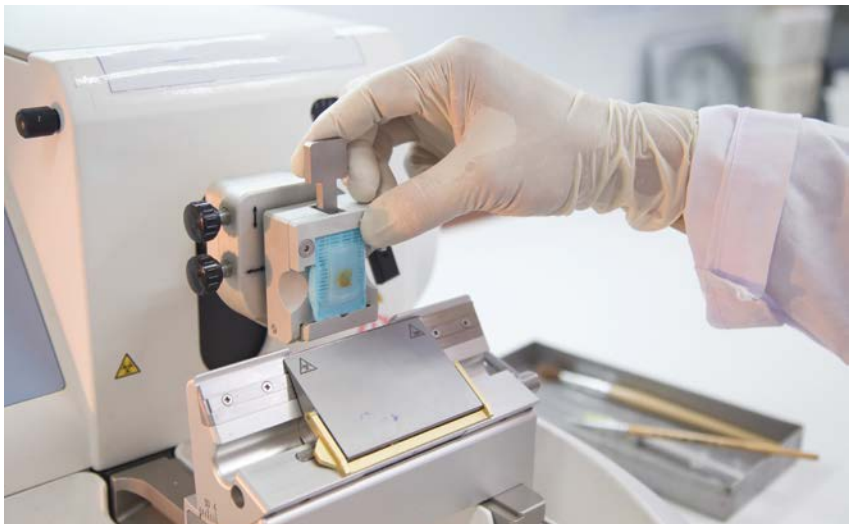
Understanding this inflammatory condition of the muscles that causes weakness.

Inclusion body myositis (IBM) is a progressive muscle disorder characterised by muscle inflammation, weakness, and atrophy (wasting). It is a type of inflammatory myopathy.

The cause of IBM is unknown. The underlying cause of IBM is poorly understood and likely involves the interaction of genetic, immune-related, and environmental factors. It is thought that some people may have a genetic predisposition to developing IBM, but the condition itself is not inherited. More males than females are affected.

Because of the inflammation associated with IBM, some doctors think the disease is a form of autoimmune disorder. In this kind of disorder, the body's immune system goes awry and attacks its own tissues – in this case, the muscles. However people with IBM don't respond at all to immunotherapy, which would normally be helpful in an autoimmune disorder. Some experts have linked IBM to infection by a virus that has yet to be identified. Other researchers believe that the primary problem in IBM is an age-related inability of the muscle to deal with destructive chemicals.

IBM develops in adulthood, usually after age 50. The symptoms and rate of progression vary from person to person. In IBM, the onset of muscle weakness usually is gradual, occurring over months or years and is asymmetrical. Falling and tripping usually are the first noticeable symptoms. For other people, IBM begins with weakness in the hands. People with IBM may have:



*IBM develops in adulthood, usually after age 50. The symptoms and rate of progression vary from person to person.*

- Difficulty with gripping, pinching, and buttoning
- Weakness of the wrist and finger muscles
- Atrophy (shrinking or wasting) of the muscles of the forearms
- Weakness and visible wasting of the quadriceps muscles (the large muscles on the front part of the thighs)
- Weakness of the lower leg muscles, below the knees

Muscle cramping and pain are uncommon, but some people with IBM do experience this. Most people with IBM progress to disability over a period of years. In general, the older a person is when IBM begins, the more rapid the progression of the condition. Most people need assistance with basic daily activities within 15 years, and some people will need to use a wheelchair. Lifespan is normal, but severe complications (e.g. aspiration pneumonia, bad falls) can lead to loss of life.

*In general, the older a person is when IBM begins, the more rapid the progression of the condition.*

Usually a muscle biopsy is required to diagnose IBM. After giving an anaesthetic, a doctor takes a sample of tissue from one of the affected muscles to be looked at in a laboratory.

When viewed under the microscope, the muscle cells of persons with IBM contain vacuoles (rounded empty spaces). Within the vacuoles, there are usually abnormal clumps of several proteins including one called amyloid. The protein clumps, or inclusion bodies, give IBM its name. This is the hallmark of IBM.

There is currently no cure for IBM. The primary goal of management is to optimise muscle strength and function. Management may include exercise, fall prevention, physiotherapy, occupational therapy, and speech therapy (for dysphagia).

The NZ NMD Registry currently has 40 people enrolled who have IBM. There have been two enquiries from researchers interested in IBM over the past six months and it is likely more will follow. Contact the registry curator registry@mda.org.nz or speak to the MDANZ fieldworker about enrolling on the NZ NMD Registry.

# The power of planning

Receiving an IBM diagnosis was a huge shock for Julie Smith, but she's discovered that with planning and help, she can do many things.

I was a primary school teacher, keen sportswoman and have always led a busy live with family and friends. When diagnosed with IBM in the year 2000 at the age of 59, I was shattered to hear there was no cure or effective treatment for this condition, and in 15 years I would be in a wheelchair!

The first sign of a problem was that I needed to use my hands to help me get up from a chair due to weak quad muscles. To begin with, IBM did not affect my life at all. But gradually, I found difficulty in doing certain things. I stopped playing tennis and took up golf, as I could still walk easily. I played until 2009 when it became unsafe to walk on uneven ground.

I continued to do normal things with the help of disability aids such as a high firm cushion, car fitted with hand controls, raised toilet seat, higher chair at home, and installed a lift and made adjustments to the bathroom. Many good ideas came from my IBM support group.

My muscle strength slowly deteriorated over the years and I have had many falls. I have broken a total of 14 bones - mostly in my legs and ribs. My worst accident was in Paris at the French Tennis Open in 2008. When exiting the Roland Garros Stadium in dim light, I failed to see a step while walking with my stick. I fell and the result was a broken femur

Julie Smith

and three weeks in a Paris hospital where a rod and five screws were inserted in my leg. During this time I used my best school French which created many hilarious situations. The French nurses could only speak a little English and I was asked, "Why do you speak English if you come from New Zealand?" Luckily my doctor spoke good English.

About five years ago my swallowing deteriorated and I could only eat soft foods and liquid. So I ate plenty of soup, yoghurt, custard, mashed vegetables, fish and chicken. The only biscuit I could eat easily was a toffee pop which I loved as a treat.

Early last year, I kept getting chest infections and losing weight. This was because food was getting into my lungs. It was now time for a PEG tube to be inserted into my stomach. I no longer eat any normal food. Overnight I have 1000mls of liquid food pumped

Julie loves visiting her grandchildren in Australia.

*I can't drive now so my husband is my chauffeur - but he doesn't appreciate it when I want to go clothes shopping!*

into my stomach while I sleep. This works really well and during the day I have water and medication through the same tube. My husband carries out my feeding programme and he is now a real expert.

For about eight years I used crutches to walk everywhere, but last year this changed because I felt unsafe. Now I use a walker inside our home and a wheelchair when I go out. I also have a power chair to use independently. I can't drive now so my husband is my chauffeur - but he doesn't appreciate it when I want to go clothes shopping!

Every year we go to Australia to visit our two daughters and their families. In May we took my wheelchair and hired a walker and a cough assist machine. MDANZ were very helpful in organising this machine and the liquid food for our visit. Our daughter already stores a raised toilet seat and a special chair for our visits.

We went on many outings and my daughters took me clothes shopping - my husband was delighted! Our 9-year-old grandson loved pushing his 5-year-old sisters around in my wheelchair. Great screams of hilarity! And at the pools and playgrounds it was, "Look Gran," and "Watch me Gran."

Our 20-year-old granddaughter was getting a new puppy so we visited animal shops to buy puppy essentials and toys. When it arrived the darling puppy shredded the book Grandad was reading. Three weeks at 25 degrees in Queensland was wonderful. It was certainly worth all the effort and organisation to holiday with our family.

I continued part-time teaching until four years ago. Now I have a carer who helps me shower daily. My arms and hands have become much weaker, but I can still write with two hands holding a pen. Each week I go to an exercise class at a hydrotherapy pool and also play Bridge at the local Bridge club. I have many visits from friends and extended family and enjoy outings with them. I try to keep active as much as I can and keep enjoying social activities. With planning and help I can do many things.

## *Support us!*

Any donation, big or small makes a difference. Donations of $5 or more are tax deductible.

**Call:** 0900 426 93 to make an automatic $15 donation.

**Online:** Donate any amount securely online. **www.mda.org.nz**

**Post:** Make a donation by post. Our postal address is: PO Box 12063, Penrose, Auckland, 1642

**Bequests:** You can create a lasting difference through making a bequest. Contact us or visit our website for information on how to include MDA as part of your will.

*Thank you. We greatly appreciate your support.*

# The times they are a changin' for neurogenetic conditions…

DR. RICHARD ROXBURGH

Watch this space!

We have just received news that we have been selected as a site for an international Huntington's disease study trialing a medication that interferes with the production of huntingtin protein. This potential treatment is similar to the drug, Spinraza, now being used around the world for treating spinal muscular atrophy. Before we initiate that trial we are about to bring a study home from Australia to New Zealand for Pompe's disease. Through the New Zealand Neuromuscular Disease Registry we have several young New Zealand patients involved in trials for Duchenne muscular dystrophy and discussions are underway regarding a potential trial in IBM. More New Zealanders with neurogenetic conditions than ever before are becoming involved in research.

*More New Zealanders with neurogenetic conditions than ever before are becoming involved in research.*

Overseas, a big study showing the efficacy of cognitive behavioural therapy in myotonic dystrophy has shown us that if you believe that you can do more, it turns out you can!!

I was at a Friedreich's ataxia meeting in Melbourne last week and there are several promising trials coming along.

I am confident that the issue soon won't be that we won't have potential treatments, but how are we going to manage with all the studies that are coming to our shores, and then how are we going to provide the medical teams to provide the care?

The problem is that until about 15 years ago, the specialty of neurology was mostly about diagnosis and not treatment. Suddenly, there will be treatments and we as a service are struggling to know how to cope. The best current examples of this are in Multiple Sclerosis and Stroke, but I feel that there is a tidal wave of treatments beginning to swell for neurogenetic conditions.

Exciting but challenging times!

*Dr. Richard Roxburgh FRACP PhD is a Consultant Neurologist at Auckland Hospital and Associate Professor at the University of Auckland's Faculty of Medical and Health Science.*

Ask the **PANEL**


# Understanding the new carer support subsidy

DYMPNA MULROY

Q: Can you explain more about the I Choose carer subsidy and what the changes will mean for me?

A: The Carer Support subsidy reimburses some of the costs of supporting the person you care for. The Ministry of Health (MoH) is changing the Carer Support Subsidy for DSS eligible clients with a more flexible type of respite support called I Choose. This will be available from 1st December.

## What is I Choose?

I Choose is a new type of disability support that can be used for 'respite' (having a break from caring for someone with disability). I Choose is money that will be paid into an eligible family's bank account once or twice per year. Families will be able to use the money to buy the respite supports and services that suit them best as long as it gives them a break from their caring role. It will make it easier for carers to have a break from their caring responsibilities.

The key changes between Carer Support and I Choose are:

- you will be paid all the money you are allocated by your NASC into your bank account
- there will be no more Carer Support claim forms to fill in and post
- you can work when using I Choose if you want to
- the break you take can be any length you like (not a half day or full day like with Carer Support)
- you and the person or organisation providing the respite support or service can agree how much the support will cost (it doesn't have to be $38 or $76 like Carer Support)

***The break you take can be any length you like (not a half day or full day like with Carer Support).***

## Who can get I Choose?

To get I Choose, you will need to be the full-time carer of a person with a disability (who is eligible for Disability Support Services funding) and have an identified need for respite.

Most people who get disability Carer Support will be able to get I Choose instead.

If you don't have Carer Support or any other type of respite support, you will need to have an assessment with a Needs Assessment and Service Coordination service (NASC) to find out if you can get I Choose.

To be eligible for Disability Support Services funding, the disabled person in your family will have a physical, intellectual, or sensory disability or autism (or a combination of these) which:

- is likely to continue for at least 6 months
- limits their ability to function independently, to the extent that ongoing support is required.

## How do we get I Choose?

From 1 December 2018, your NASC will talk to you about changing from Carer Support to I Choose at your next review or reassessment. Most people who have disability Carer Support will change to I Choose during 2019. If your NASC allocates you I Choose, you will get a letter in the mail from the Ministry of Health about what you need to do to set up the payment. Please make sure that your NASC has your current postal address.

## What will I be able to buy with I Choose?

I Choose aims to give you as much flexibility as possible over what you can buy. You can spend I Choose on any support or service that helps you have a break from caring for person with a disability. It's ok to think 'outside the box' about your respite options – some ideas include:

- asking a friend to hang out with

*It's ok to think 'outside the box' about your respite options.*

the person with a disability in your home while you go out

- paying for a fun activity for the person with a disability
- paying the travel costs for a friend or family member to stay and help out in the school holidays
- employing or engaging a support worker to help you have a break.

## Who can I pay with my flexible respite budget?

You will be able to use I Choose to pay people or organisations who help you to have a break from the caring role. This can include non-family relief carers who live at the same address as the disabled person such as an au pair or flatmate.

You will not be able to pay or compensate:

- family members who are living with the disabled person
- the disabled person's parent or spouse

## Will my flexible respite budget affect my income support or other benefit eligibility?

No. The disability support payments you get from MoH are not income. This means I Choose won't be counted as income when determining your eligibility for a main benefit or supplementary or hardship assistance from Work and Income.

## Will I Choose be available for people who get DHB funded Carer Support (for mental health, long term chronic conditions or age-related support needs)?

There is no change for Carer Support that is funded by District Health Boards (DHBs) at this stage. DHBs are considering whether I Choose or something similar will work for people that get Carer Support for support needs related to mental health, long term conditions or age.

## How can I find out more?

Please ring 0800 855 066 (select option 4 Disability Support Services) to talk to a person who can help you, or email respitestrategy@moh.govt.nz.

*Dympna Mulroy is the new Member Services Manager at MDANZ.*

# Self care and resilience during busy times

## *The end of the year can be overwhelming*

The P.E.A.K model is a helpful way to maintain resilience.

*Self care is a word for the set of healthy behaviours and attitudes we deliberately employ to look after our own well being leading to a physical and mental 'stress buffer'.*

The word **P.E.A.K.** is an easy-to-recall acronym for a set of self-care strategies you can use to maintain resilience. **PEAK stands for Prepare, Energise, Ask and Know.**

## Prepare

### Prepare physically and organise your self

- Inform your social network, spouse and family about the peak times coming up and how this might change your timetable, availability and energy
- Plan around the possible timetabling challenges for other demands in your life – you might need to put some things on hold or rearrange things like childcare and social events
- Ensure a good sleep pattern by going to bed on time and the same time each night
- Keep up physical activity – walking and general exercise are fantastic for resilience
- Organise and plan meals, so you avoid skipping meals or eating junk, therefore not missing out on good nutrition.

### Prepare mentally with realistic expectations

- Because what we think greatly determines how we feel and react – it's best to give yourself some mental preparation in the form of 'positive self-talk' to combat negative thinking about peak times
- Use self-talk statements like: *"Yep, it's a busy time again and I can expect to be tired at times, but I have some strategies in place to keep my energy levels up.."*
- Develop a personal motto which brings your focus back to the positive work and the enjoyment of the job, such as *"I really like this work and the benefits it brings me" or "No matter what comes my way, I can make my day".*

## Energise

### Energise physically each day and throughout the day

- A lot of people fail to recognise that frustration can stem from actually being hungry, tired or dehydrated – and not really about the situation in front of you at all
- Eat before you go on your shift and eat your lunch/ dinner in your break
- Drink more water and less Red Bull or coffee!
- Use a quick deep breathing or relaxation techniques in a quiet space to relax
- Use a quick energiser to stimulate the endorphins (positive hormones) – eg stretching, a quick walk, singing (quietly if needed), shaking our your arms and legs. Simple but effective.
- Determine your own practical 'stress busters' or 'energisers' that you can use each day and let your colleagues know, so they can remind you to use them

### Energise mentally each day by using your own positive focal point

- You can make your own day by choosing and using a positive focal point whether it be a statement, a memory, an attitude or person
- Recall all the positive situations, the successful jobs and fun with colleagues
- Use your personal positive motto OR 'borrow' one you like from someone else

- Think about a positive event or person or scenario – even for a few seconds – to re-set your mind
- Try a quick meditation or visualisation technique (many people already know how to do this AND you can learn then by attending a personal consultation with EAP)

## Ask

**Ask questions outwards to others to open up good communication**

- When we're feeling tired or stressed, we tend to restrict our communication rather than slow down, ask questions and listen more. This can lead to misunderstandings.
- Ask questions to determine others' expectations of you – this takes out the guess work and avoids potentially awkward encounters
- Just by clarifying what can be achieved, and asking colleagues and managers about how you can jointly manage the load, can lead to a reduction in pressure, as you determine realistic expectation together
- Ask if you can help others – always a winner – If you're in doubt or stuck in a challenging conversation, gently ask what the main issue, concern or need is. AND ask *'how can I help you' and 'what would you like me to do'.* This tends to defuse frustration and lead towards some better information that you can actually deal with
- Ask for help and support when you need it.

**Ask questions inwards to yourself to maintain self-awareness in the here-and-now**

- One of the best ways of avoiding stress and conflict with colleagues is to develop immediate self-awareness of your own frustration levels and enable yourself to act more deliberately and positively
- Asking questions of yourself can, in itself, be a good 'stress buster' as it can lead to a broader and less pressured perspective. The answer usually comes into mind and based on your past experience.
- "How did I handle this type of situation last time?" *"What can I realistically achieve before the end of my shift now?" "Am I reacting strongly to this colleague or client today because of another reason, like being tired"? "What's the best way I can manage this now, that will lead to a good outcome – rather that what I'd really like to say.." "How will my actions right now likely impact on the other person – and do I want that?"*
- Ask yourself questions to form a check-list approach for daily resilience. For example: *"Have I had enough to eat and drink today? "Have I had a good break?"*
- Ask yourself honestly – if you might need to brush up on your resilience skills – taking action to join a course or gain coaching or counselling can be one of the most rewarding steps you take in life.

## Know

**Know your physical limits so you stop short of the edge**

1. Accept that you will have limits to your time, energy, patience AND skill-sets and this may even be changeable depending on what's happening in your life
2. Determine your own healthy limits and communicate these to your close network. You can take a lot of pressure out of setting limits with others by simply and unashamedly telling them what you can manage and what you can't (right now)
3. Some limits you might want to monitor in case you run the risk of stress are: the amount of sleep you need; the amount of extra-curricular activities you can squeeze in; the amount of time you need personally to re-charge; how long you can go without using your entitled leave due to you; how many extra jobs you can handle for others at work on top of your usual tasks.

**Know your own early warning signs of stress so you stop short of boiling point**

Everyone has early warning signs of stress and if you catch them early you can use a practical strategy to maintain your balance both in terms of wellbeing and for interactions with others:

Resilience can be developed and stress can be managed. You can make it happen by determining your own **PEAK Plan**, by selecting your set of strategies - **record them, talk about them and do them**!

Reproduced with thanks to EAPworks (www.eapworks.co.nz) and their Austrailan partner Assure Programmes Australia.

Muscular Dystrophy New Zealand

# About us

MDANZ is a trusted source of specialist information and provides a range of free services and practical support for individuals, families and whānau with lived experience of rare neuromuscular conditions.

The Muscular Dystrophy Association of New Zealand Inc., commonly known as MDANZ, began in the late 1950. Since then MDANZ has broadened its scope to support many other neuromuscular conditions. We are proud to have Judy Bailey and Dame Susan Devoy as our longstanding patrons.

Our unique governance structure ensures leadership of the organisation by individuals and family members with lived experience of a neuromuscular condition. We have four regional branches that are supported by the National Office based in Auckland.

We want New Zealanders with lived experience of neuromuscular conditions to experience freedom of choice in a responsive society.

To achieve this mission, we provide;

- Free information and advice, through our website, an 0800 info line and in paper booklet form
- A nationwide fieldworker service for personalised support
- Free loan of resources, such library books, recreational beach chairs and cough assist machines
- Funded support for counselling
- Discretionary funding for life enhancing resources not covered by government
- A high quality quarterly magazine to inform and inspire our membership and broader communities of support
- Funding for neuromuscular research and a mechanism to help New Zealanders to access clinical trials and new treatments
- Education workshops for members, health professionals, schools and others
- Advocacy and lobbying at a community or national level
- A platform for support groups and peer to peer networking

MDANZ is a registered charity and relies almost entirely on donations from the public, trusts and other businesses/ organisations to continue its work in the community.

## Our core team

*Ronelle Baker*
Chief Executive

*Miriam Rodrigues*
Programme and Service Advisor

*Brian Hadley*
Accountant and Business Manager

*Dympna Mulroy*
Member Services Manager



See more about our team at www.mda.org.nz

### Northern Branch


Fieldworkers: *Darian Smith and Rachel Woodworth*
Office Manager: *Denise Ganley*
Ph: 09 415 5682 or 0800 636 787
Email: support@mdn.org.nz

### Central Branch

Fieldworkers: *Penny Piper*
Ph: 0800 886 626
Email: info@mda.org.nz

### Canterbury Branch


Fieldworkers: *Paul Graham and Marty Price*
Ph: 03 377 8010 or 0800 463 222
Email: mdacanty@xtra.co.nz

### Southern Branch

Fieldworker: *Jo Smith*
Ph: 0800 800 337
Email: joanne@mda.org.nz

## Council Representatives

If you want issues brought to National Council meetings, talk to your branch representative. They have the responsibility to raise your issues at National Council meetings and to make sure you are heard. Your branch representatives and their contact details are as follows:

### Northern Branch

Michael Schneider. Ph: 021 851 747
Email: spider@spider.co.nz

### Central Branch

Bernadette Ingham. Ph: 027 600 3868
Email: members.central@mda.nz

### Southern Branch

Andrew Willetts. Ph: 027 371 7573
Email: andrewwilletts72@gmail.com

### Canterbury Branch

Rebecca Poad.
Email: rebeccapoad@yahoo.com

# Conditions covered by MDANZ

### Muscular Dystrophies:

Becker Muscular Dystrophy

Congenital Muscular Dystrophies and Congenital Myopathies

Distal Muscular Dystrophy

Duchenne Muscular Dystrophy

Emery-Dreifuss Muscular Dystrophy

Facioscapulohumeral Muscular Dystrophy

Limb-Girdle Muscular Dystrophy

Manifesting carrier of Muscular Dystrophy

Myotonic Dystrophy

Oculopharyngeal Muscular Dystrophy

### Diseases of the Motor Neurons:

Spinal Bulbar Muscular Atrophy (Kennedy's Disease and X-Linked SBMA)

Spinal Muscular Atrophy - all types including Type 1 Infantile Progressive Spinal Muscular Atrophy (also known as Werdnig Hoffman Disease)

Type 2 Intermediate Spinal Muscular Atrophy

Type 3 Juvenile Spinal Muscular Atrophy (Kugelberg Welander Disease)

Type 4 Adult Spinal Muscular Atrophy

### Hereditary Spastic Paraplegias (HSP)

*- all types:*

Also called Familial Spastic Paraparesis

### Leucodystrophies

- all types.

### Metabolic Diseases of muscle *- all types including:*

Acid Maltase Deficiency (also known as Pompe's Disease)

Debrancher Enzyme Deficiency (also known as Cori's or Forbes' Disease)

Mitochondrial Myopathy (including MELAS, MERRF, NARP and MIDD)

Phosphofructokinase Deficiency (also known as Tarui's Disease)

Phosphorylase Deficiency (also known as McArdle's Disease)

### Diseases of the Peripheral Nerve:

Charcot-Marie-Tooth Disease (CMT) (Hereditary Motor and Sensory Neuropathy) - all types

Dejerine-Sottas Disease (CMT Type 3)

Hereditary Sensory Neuropathy

### Inflammatory Myopathies:

Dermatomyositis

Inclusion Body Myositis

Polymyositis

### Diseases of the Neuromuscular Junction:

Congenital Myasthenic Syndrome

Lambert-Eaton Syndrome

Myasthenia Gravis

### Myopathies *- all types:*

Andersen-Tawil syndrome

Central Core Disease

GNE Myopathy

Hyperthyroid Myopathy

Hypothyroid Myopathy

Myofibrillar myopathy

Myotonia Congenita (Two forms: Thomsen's and Becker's Disease)

Myotubular Myopathy

Nemaline Myopathy

Paramyotonia Congenita

Periodic Paralysis

### Inherited Ataxias:

CANVAS

Friedreich Ataxia (FA)

Spinocerebellar Ataxia (SCA)

### Neurocutaneous Syndromes *- conditions affecting the brain and the skin:*

Central Cavernous Hemangioma

Neurofibromatosis Type 1

Neurofibromatosis Type 2

Schwannamatosis

Tuberous Sclerosis

Von Hippel Lindau Syndrome

*Should you have a query regarding a condition not listed please contact us on 0800 800 337 or email info@mda.org.nz*

EVERY DAY. EVERY WHERE

# TA Junior offers more freedom...

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Fits lower under standard tables and easier in small crowded spaces.

> Minimum seat plate height 38cm/15”

> Seat base can grow with child

## AT HOME

Comes standard with hi-low elevate & tilt function. Sit low, sit high.

> Seatplate from floor 38cm/15” to 68cm/26.5”

> Tilt to 45°

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> Crash test approved for occupant to remain in chair in the vehicle

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