InTouch

KIA NOHO TATA Inclusive | Inspiring | Informative. Spring 18 Issue 100

The power of pets

Providing happier, healthier lives

The Freedom issue

Getting behind our annual campaign

The power of community

Joining together for progress

Sneezing season

Coping with hayfever

Muscular Dystrophy New Zealand

InTouch

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Catching up with Trevor

Kia ora everyone.

In the first five weeks of performing the role of Chairperson for National Council, I have been surprised and also encouraged. It has been a busy time, and with change comes opportunities for new learning. I have definitely gained a different perspective about our organisation in this short time.

I have always been passionate about MDANZ and my own efforts as Northern Branch Chairperson have been focussed on doing everything I can to ensure support is available for our members. What has become apparent to me, is that the organisation itself is a very important vehicle for achieving this support.

The organisation does need to be well run, by skilled people. I have been encouraged by the loyalty and dedication by the team of paid staff and all of the volunteers on governance committees, who are the machinery that keeps everything working. I appreciate the hard work going on at all levels of the organisation.

But as a whole, MDANZ is vulnerable. Competitive conditions for fundraising, stretched human resources and tired committee members are a risk for our sustainability. During my term, I am committed to ensuring stability and will work hard to nurture stronger relationships between our stakeholder groups, and as with Ken Green the former Chairperson, I will ensure we keep our focus on our members.

If we take small steps together, we will manage the current changes more effectively and find a new way forward for the future.

Trevor Jenkin
National Council Chairperson

Our new chairperson

We are sorry to inform our membership that Ken Green has chosen to discontinue in his role as National Council Chairperson and as Vice Chairperson, Trevor Jenkin will be performing the role of Chairperson of the National Council until the next AGM.

Trevor was elected for a two year term as Vice Chairperson in April 2017 following a five year period of service as Chairperson for the Northern Branch, where he was often the Branch Representative on National Council.

He resides in Auckland with his wife Joy and both are very passionate about MDANZ, its members and services. Their son who had Duchenne muscular dystrophy, sadly passed away in November 2015 at the age of 17. Trevor is a former civil engineer and is now self-employed with a successful business. He enjoys interaction with members and fostering relationships so that members may share information and learn from each other.

In touch with Ronelle

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

Nelson Mandela once said, "…to be free is not merely to cast off one's chains, but to live in a way that respects and enhances the freedom of others."

I believe this is the very essence of our organisational mission and our awareness campaign in September, which invites all New Zealanders to reflect on what freedom means to our families and whānau living with neuromuscular conditions, and help us create a more responsive society that enhances our freedom.

But before we get there, August is SMA awareness month and this community and those who support them deserve a big shout out! Our SMA Member's Reference Group is developing a sense of identity and cohesion, setting some exciting goals to achieve before the year ends. This is a group that is reciprocal, purposeful, and fosters collective learning.

On page 16 Reference Group member Fiona Tolich shares passionate insight following attendance at the recent Cure SMA Conference held in Dallas Texas. Other contributors this issue include Raymond Mok, who tells us how a sip ventilator provided by MDANZ has given him greater freedom in life, and guest column writer Liz Church, who talks about her new experiences receiving personalised support at home.

A number of members have also contributed to our feature article about the power of pets in their lives. I thank my parents because I always had pets around growing up. And even though my children have allergies and eczema, I've made sure they have pets in their lives too - because I believe that pets help children develop empathy, and there is nothing like the unconditional love of an animal to make a person feel better on a bad day. Small things can make a big difference and my Chihuahua/Papillon cross 'Paco' did that for me, at a time when transitioning to a power chair felt emotionally hard. So here's to warm fuzzy friends everywhere!

Hei konā rā, Bye for now.

Ronelle

Ronelle Baker
Chief Executive

P.S. With spring just around the corner, it's time to think about getting back into the garden. Our friends at Amazing Iris Garden have come up with a great fundraising offer. Go online to www.irisgarden.co.nz and purchase of pack of differently coloured historic irises for $38, and half the proceeds will be donated to MDANZ.

My lovely dog Paco.

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.

A cup of tea and a catch up with … Melanie Kerr

Each issue we introduce a MDANZ team member:

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

I started in May this year and am the Executive Assistant to our Chief Executive.

What qualifies as a great day at work for you?

A great day is when I get to tick boxes and know indirectly that the work I do makes a difference.

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

Equal Opportunities and inclusiveness.

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

Variety is the spice of life, so a little bit of everything, savoury and sweet.

What are you passionate about?

My family, animals and the environment.

Save the date

Next year will be an exciting year for MDANZ! Planning is well underway for the 2019 Neuromuscular Conference to be held at Waipuna Hotel and Conference Centre 1-2 August in Auckland, and offers an exciting programme that aims to improve the health and wellbeing of individuals, families and whānau living with neuromuscular conditions, through research, education and collaborative practice. Themed *Freedom beyond limits*, the conference will combine a milestone event as the Muscular Dystrophy Association of New Zealand (MDANZ) celebrates its 60th Jubilee.

Keep an eye on **www.mda2019.org.nz** for further developments.

Your thoughts on the NZ Carers Strategy

Thank you to all of the members who responded to our survey about the NZ Carers Strategy.

A total of 276 members responded to the survey, however many people skipped the questions and did not provide detailed feedback. 57 percent of respondents were individuals with neuromuscular conditions, 17 percent were parents or guardians providing informal care and 14 percent were full-time carers.

More than half of respondents skipped the question about the Carer Alliance priorities. However, of those that answered the question, the following priorities were important to participants: 73 percent Carer Support, 58 percent payment for family carers, 35 percent respite and 25 percent continence products and support for continence issues.

The overwhelming feedback from our members is that acknowledgment of family carers (including parents and spouses) and ability to have them receive pay is still an important issue for New Zealand.

In addition, caring for carers is essential. Suggestions included continued education for carers, promoting the current support available for carers, introducing new support – such as subsidising their GP visits and vaccinations, ensuring flexible respite options for carers to have breaks, and provision of appropriate modifications, products and equipment that enable caring to take place at home.

MDANZ is a member of the Carers Alliance. You can read more about the work of the Alliance at http://carers.net.nz/nz-carers-alliance/

Our bid to join the Disabled Person's Organisation (DPO) Coalition

MDANZ has applied to become a recognised DPO in Aotearoa New Zealand. If successful, we will join the DPO Coalition and take a seat at the table with government. The DPO Coalition is responsible for representing the voice of its members and ensuring disability-led monitoring of the Convention on the Rights of Persons with Disabilities (CRPD), NZ Disability Strategy and NZ Disability Action Plan.

Happy Birthday *In Touch*!

Hip, hip, hooray we are 100! We're very proud to present you with the 100th issue of *In Touch* magazine. What began as a newsletter to members, grew into a fully fledged magazine in 1991, with the very first issue on glossy paper.

Since then, the look and feel of the magazine has changed considerably, but one thing that has stayed the same is it's a trusted source of information, and a way for members to connect with each other. Here's to another 100.

Northern's midwinter lunch was a great way to escape the cold.

From the branches

Northern

Members enjoyed escaping the cold and catching up at a series of mid-winter lunches in Auckland, Rotorua and Whangarei.

Wellington

Wellington has closed its branch office in Petone and is moving towards a more mobile, digitally connected service. You can keep in touch via phone 0800 886 626 or email members.central@mda.nz.

The branch farewells Annelize Steyn from the Chairperson role and welcomes Tristram Ingham to the role. Bernadette Ingham will perform the branch rep on National Council.

Canterbury

A big thank you to Sammy Metherell who took part in the University of Canterbury Student Association's Charity Fight Night boxing tournament to raise money for MDANZ. It was a great way to raise both funds and awareness.

The branch farewells Andrew Munro as Chairperson and welcomes Rebecca Poad, who will also takes up the branch rep role on National Council.

Southern

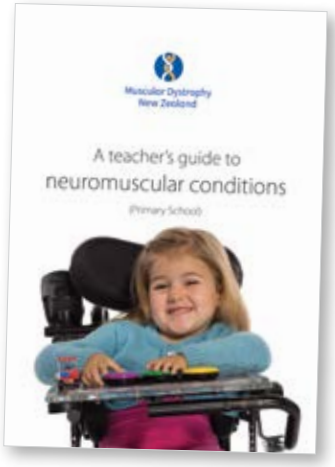
A great time was had by the children who attended the Lego group in Oamaru and the event will definitely be repeated. Thanks to Fresh Choice Dunedin for the recent support.

Fibre is here

National Office now has ultra-fast broadband installed. This is a great investment into our infrastructure and makes connectivity better for our remote workers. If you are in the area and want to make use of our accessible facilities and wi-fi, please drop by. Our place is your place.

On the look out for Dukies

Are you ready to take your life to the next level and set goals to challenge yourself? If you are between 14 and 24 years of age and want to find out more about our Duke of Edinburgh Hillary Award programme, get in touch with Marty Price; Email marty@mda.org.nz; Phone 027 444 6681



New look resources for schools

We've updated our resources for schools, so that teachers have a better understanding of what living with a neuromuscular condition means for kids, and can help make sure they get the most out of their education.

We worked with parents, asking what they would like teachers to know, and also asked some children to share their experiences in the classroom and playground.

The booklet is aimed at primary schools, and we will be following it up with a secondary school version shortly. If you'd like a copy for your school, you can download it from our website www.mda.org.nz, or email info@mda.org.nz and we will post you a copy.

Medication use in myotonic dystrophy

Post graduate scholar reveals new findings about medication use for people living with myotonic dystrophy in New Zealand.

Post graduate scholarship recipient Miriam Hanna is well on her way to completing her research into the pharmacological and non-pharmacological treatment of people living with myotonic dystrophy (DM) in New Zealand. A qualified Pharmacist, Miriam is completing her MPhil through AUT University and is completing a cross sectional study of current treatment for DM in NZ. She has utilised the data on 213 individuals with a confirmed diagnosis of DM identified through a nationwide population-based prevalence study, MD-Prev.

Myotonic dystrophy is the most common form of muscular dystrophy and while progression is usually considered to be slow, DM is extremely variable and associated with a large number of comorbidities. Medication use, as well as herbal and vitamin supplements for this population are therefore of interest to the neuromuscular community.

This study used statistical methods such as t-tests and chi square tests to explore differences in those using pharmacological or non-pharmacological treatments. A logistic regression explored the factors that significantly predict treatment use.

The participant group (sample) was predominantly adults (96 percent), of European ethnicity (92 percent), diagnosed with DM1 (94 percent). Results showed that 57 percent were taking prescription medications, with some participants taking up to 13 medications. The majority however (80 percent) were taking up to four different medications.

A total of 120 different prescription medicines were identified and grouped according to the WHO ATC/DDD classification. Only eight percent of participants were taking nutritional supplements, but over one third (35 percent) were using herbal supplements or vitamins. Nine percent were paying for non-funded medication.

The overall impressions from Miriam's study are that medication use in DM is high, with many associated comorbidities and symptoms treated simultaneously. Predictors of medication use include access to neurology, hospitalisation and comorbidities, reflecting increased severity of DM. Interestingly pain and fatigue were not correlated with medication use. There was also no link between medication use and use of alternative therapies.

Key recommendations from this study include improved access to neurology/neuromuscular clinics, annual medication reviews through community pharmacy to assess suitability, adherence and compliance, and improved funded access to medicines that could be useful for DM.

Grant funding round open

Neuromuscular Research New Zealand (NRFT) is once again accepting applications for funding research relevant to New Zealanders living with neuromuscular conditions.

Grants of up to $30,000 will be considered, and as with previous years, up to two postgraduate student scholarships of $5000 will be considered.

Research of a preliminary nature with the intention of developing further proposals for substantial financial support from elsewhere is encouraged.

Closing date for applications is 30th September 2018 and applicants will be advised of the outcome by 1st December 2018.

For more information and an application form, please email Miriam Rodrigues, miriam@mda.org.nz or go to the MDANZ website www.mda.org.nz/Our-Research/Apply-for-Funding for information and to download an application form.

Freedom diary dates

7th Sept - DMD Awareness DAY and campaign launch

Morning tea for MD, at Northern Branch offices, 3 William Laurie Place Albany, Auckland.

8th Sept - Fly for Freedom kite making family day

Mt Albert Senior Citizens Hall (beside Rocket Park) , 751 New North Road, St Lukes, Auckland

Check Facebook and our website for other branch locations across the country.

9th Sept - Craft stall Carterton Daffodil festival

Celebrate spring at the Carterton daffodil festival and support the Wellington Branch craft stall.

14th Sept - Freedom Street Appeal

Street collections will take place all around the country. Collections may take place on other days.

15-16 Sept - FA Family Forum

To be held by FARA NZ in collaboration with MDANZ at the Brentwood Hotel Conference Centre, 16 Kemp Street, Kilbirnie, Wellington. The forum will bring families together and provide an update on what is happening in NZ for people with FA. Contact Dianne Boon for information or to RSVP diboon27@gmail.com.

22 Sept - SMA Family Day

MDANZ National Office, Penrose, Auckland on Saturday 22 September. Travel assistance is available. Contact Natalie Brunzel to RSVP or find out more natalie@mda.org.nz.

29th Sept - Casino night in the Bay

A Casino evening will be held in the Hawkes Bay. Tickets are $25. Members are welcome to volunteer to help out and watch the fun, or donate food items for supper. Contact Penny Piper penny@mda.org.nz for information.

29th Sept – Member Education Session

To be held from 1-4pm at the Commodore Airport Hotel, Memorial Ave, Christchurch. Join in for afternoon tea and hear from a range of speakers. Contact Gemma Foulds gemma@mda.org.nz for information or to RSVP

30 Sept - LGMD awareness day

Join the northern team in a walk around the Hatea loop at Whangarei basin, followed by a Freedom lunch. Contact Denise Ganley denise@mdn.org.nz for information.

6th Oct Casino night in Dunedin

A Casino evening is planned for 6th October. Contact Jo Smith joanne@mda.org.nz for tickets or to get involved.

September is also CMT Awareness month! N

Our Freedom Campaign

Celebrate Freedom with the Muscular Dystrophy Association of New Zealand (MDANZ) this September, and help us to raise awareness and vital funds for local services.

We want to start a conversation amongst our members, and all New Zealanders, asking, "What does Freedom mean to you?"

For our members these may well centre on everyday freedoms that can easily be taken for granted, like driving their own vehicle, living independently in their own home, making a cup of tea, cooking a meal, to broader concepts like having a career and relationships, or parenthood.

There are many ways you can get behind MDANZ during the month of Freedom;

- Volunteer to fundraise for your local branch
- Help out as a collector for the Freedom Street Appeal
- Display one of our donation boxes in your workplace
- Like us on Facebook
- Share a picture or video talking about what freedom means to you
- Hold a morning tea for muscular dystrophy
- Join the schools around the country already holding mufti days. N

6 steps to organising a morning tea for muscular dystrophy

Some of the most successful fundraising is done peer to peer.

Holding your own morning tea is a great way for your friends and colleagues to get behind our month of Freedom and to raise the profile of MDANZ and your local branch of MDA. It doesn't have to be a huge event because every single donation makes a difference to the lives of our members.

1. **Choose your guests:** This is a great time to get together for a good old cup of tea and a catch up with your family, friends, workmates and support networks – and raise money at the same time.
2. **Confirm your date, venue and time:** There are several ways to approach raising money this way. You can have the morning tea at your home and provide the food and drinks with everyone giving a donation to MDANZ in acknowledgement of your effort.

   Or, you can agree to meet at a cafe, with everyone buying their own coffee and muffin, but still making a donation at the end.

   If you have children, consider holding your morning tea at a local park or playground. Fill a couple of picnic baskets with snacks and drinks, and pack a thermos for hot drinks (even better, choose a park that has a coffee cart).
   Your workplace is another great venue – everyone loves the chance to share morning tea together – and contribute to a great cause.
3. **Send out invitations:** The earlier you can get invitations out the better, as people's calendars get booked up quickly. We've designed some generic invitations that you can download from our website www.mda.org.nz. You will just need to add your own time, date and place. Remember, it's always a good idea to send a reminder text or email the day before.
4. **Plan your menu:** Remember, you don't have to go overboard with a wide variety of food – unless you really want to! A batch of blueberry muffins always go down a treat, or go classic with fresh scones and jam and cream (great if you're taking them into the office). For the easiest scones ever, Google lemonade scones, just a few simple ingredients and they work every time.

   If you're planning an outdoor morning tea, grazing platters work well and are easy to transport. Stock up on cheese, crackers, fruit and dips.
5. **Organise decorations:** If you want to bring a festive air to your morning tea, use the MDANZ colours of blue and yellow for napkins, tablecloths, flowers, bunting and balloons.
6. **Collect the donations:** It's up to you whether you request a specific amount, or simply ask for a donation. Get in touch with your branch and they will provide bank account details so you can deposit the amount you make, or your guests can make an online donation themselves.

Cover star Jodie Thorne and her horse Tech.
Photo credit: Rhiannon Moss, NZEquine

The power of pets

Whether they live in a kennel, a hutch, or out in the paddock, pets offer their owners a happier, healthier life.

Three members explain the bond they share with their animal friends.

Jodie and Tech

When it comes to a perfect match, you don't have to look further than Jodie Thorne and her beautiful horse Tech.

In 2010 Jodie completed a riding therapy course at Tauranga Riding for the Disabled, which reignited her childhood love of competing. In order to do so, she needed to find a very special horse.

"It took a long time to find the perfect horse due to my unique requirements and the way I ride with my FSHD," says Jodie. "After nine months of searching, I finally found Tech in Maramarua and the rest, as they say, is history! He really is my perfect horsey partner - safe, sensible and quick to learn - with a very cool personality to boot!"

Several years down the track, Jodie says Tech has improved her life in many, many ways.

"Firstly, riding is such a good form of exercise for me. I won't get strength back that I've already lost through FSHD, and as a progressive disease there are certainly things I can't do now that I could when I first bought Tech (I was walking when I first got him - now I'm in a wheelchair 99 percent of the time). But my stamina has definitely improved, and riding regularly is keeping my body supple and moving which I believe is helping to slow my progression. I really feel a difference if I can't ride for more than a week or so - I get really stiff and feel lethargic and just yuck. I try and ride three to four times a

Cover photo credit: Kaitlyn Perry Photography.

week, more if I can work it in. Riding is fantastic for the body and soul."

Tech has also given Jodie the pleasure of setting goals and competing in national competitions each year, and introduced her to a whole new community of people, including her coaches and a volunteer crew.

Jodie says spending time with Tech is a great stress reliever.

"It makes any stress or worries disappear, and gives my energy and general happiness a boost. I'm sure my family will attest to the fact I'm definitely happier when I can ride my horse! If I'm not riding, I love just chilling out in the paddock with Tech, feeding him treats or doing some fun clicker training with him - teaching him to come to a whistle or back up off just my hand gesture. He's such a character and we have a very special bond. I love him to bits!"

While he's very much at home in the competitive arena, Tech is also a much-loved family pet.

"My husband and son will go up to the paddock and give him a pat and a carrot and he's definitely a big part of the family. If he could live in my garden with our dogs I'd have him there in a flash!" says Jodie.

She wholeheartedly recommends spending time with horses to other members.

"Do it! It's such a great form of exercise, and being able to compete at Para-Equestrian Dressage competition is like the icing on the cake! If not for the exercise, do it for the social aspect. Meeting new people and making lifelong friends is all part of it. If not for the social aspect, then do it for the relationship and bond you build with such an incredible animal. Being able to safely sit on a 600kg animal and control him with my breathing, small weight shifts and small hand and leg movements is quite mind-blowing. There really is nothing like it!"

Jodie and Tech in competition mode. Photo: Rhiannon Moss, NZEquine.

Being able to safely sit on a 600kg animal and control him with my breathing, small weight shifts and small hand and leg movements is quite mind-blowing.

Dylan and Ziggy

When you're feeling a bit down, there's nothing better than the company of someone who seems to understand exactly what you're feeling.

For Dylan Schneider, that comfort comes from Ziggy, his retired racing greyhound who has been sharing his life for nearly five years.

"He likes to cuddle up close and is always there for me at bedtime. Ziggy makes me happy, and a lot less lonely, and always seems to understand when I'm feeling down", says Dylan. "I rely on him when I am sad, he makes me feel better. When I am bored, he keeps me entertained."

Dylan and Ziggy have a very special bond.

The Schneider family has had a variety of pets over the years – cats, guinea pigs, rabbits, rats, fish and tadpoles. They also share their home with Jamie, Ziggy's second cousin, who has a very different temperament.

"Ziggy is very cuddly and placid, loves people and is chilled out."

Dylan's best memory of Ziggy is of the day he first arrived.

"He jumped up into the van as if he had been living with us forever, and he rested his head on my lap."

Ziggy has brought so much to Dylan's life that he's keen to share some of the happiness around and join the SPCA and St John Outreach Therapy Pets Programme.

"I decided to do this as the social side to my Duke of Edinburgh Hillary Bronze Award. Because Ziggy is so chilled out, and loves the cuddles, I wanted to show him to the elderly residents at my 93-year-old Nana's rest home. I think it will go just great as Ziggy is so chill, and just loves people. We have had to wait until I get my new disability van that will take me in my new Levo wheelchair. Once we have the van, my Mum and I are going to do the training."

Niki and friends

Feeding time at Niki Wright-Jackman's home involves quite an extensive roll call. There are five cats named Tommy, Coco, Cheezel, Cookie and Tinkerbell, and two dogs, an Akita named Bailey, and Halo the Rottweiler.

Niki has always loved animals, and can't imagine her home without them.

"Pets teach and give unconditional love. They also teach responsibility and as I used to live alone they gave me so much pleasure when I came home each day from work. Now 20 odd years on they still give me a lot of pleasure by way of companionship. They are my family."

While Niki adores her own two dogs, she says she has a special affinity with cats. She has a very special way of communicating with them, and her 'cat talking' skills have been able to help out other pet owners. You can find out more about that at her website www.zengal.co.nz.

Not surprisingly, Niki's husband and son love pets as much as she does. She has worked hard to make sure her animals accept her son, and each other.

"Even before our son was born I allowed our dog (we only had one at the time) to go into baby's room, and once he was born, I would bring home clothes to wash and let our dog smell them, so when we bought our son home from the hospital our dog was already used to his smell. We let our dog be in the house at all times supervised, except for when our son was a baby and had tummy time, then our dog was put outside. Seven years on they are great friends. It makes me happy that our son is growing up with pets.

"The dogs have grown up with the cats, so they all seem to live in harmony but they have their specific areas. The cats have a sunroom. The dogs have a kennel and crate."

Niki has always shared her home with pets.

Cheezel the cat is one of five.

Niki takes her responsibility as a pet own very seriously, and says while they will make your life better, it's important to plan carefully before taking on the responsibility.

"I am a responsible pet owner. So even though I may not be able to do a lot physically to care for them, I ensure they are looked after. Every dog needs walking no matter what size. My husband walks our dogs, but I am able to also walk one of them at the moment. Both cats and dogs need companionship, dogs especially as they are a pack animal.

Four ways pets are good for us

They relieve stress

Cats and dogs love to be petted, and it does you both good. Stroking or cuddling a pet boosts oxytocin levels, a hormone associated with social bonding, which has also been shown to reduce anxiety and depression. Pets can also lower stress hormones (like cortisol) and lower your heart rate. Studies have found that dogs can help ease stress and loneliness for older people, and help calm high school and university students before exams.

They ward off depression

Pets keep loneliness and isolation at bay and often make us smile, even when we don't feel like it. They help keep us engaged in daily life (via their daily demands for food, attention and walks) when things feel overwhelming, making them an excellent way to ward off the blues

They make us social

Pets, especially dogs, can help you connect with other people. Studies show four-legged companions (particularly dogs that get us out of the house for daily walks) help make us friendlier and appear more approachable and trustworthy to others.

They're good for our heart

According to the US's National Centre for Health Research, blood pressure is reduced during stressful times for people with pets, as opposed to those without them. Studies have found correlations between having an animal and reduced anxiety, which, in turn, amounts to a healthier heart.

Finding my freedom

The path to confidence and courage

As we approach our annual Freedom campaign month, we're turning the spotlight on what freedom means to each of us. Raymond Mok found Freedom through finding his voice, and realising he had a powerful story to share. A positive new attitude – and the right breathing equipment – has opened up a new world of possibility.

I have been a shy person ever since primary school. I realised that the lack of confidence was holding me back from reaching my potential.

I struggled with the hesitation to say what I wanted to say, and to ask for help, so I wanted to become a more confident and better communicator. Through participating and speaking at Toastmasters, I learnt public speaking. I have gained the courage to share my personal story in front of people I know, and those I don't know.

At Toastmasters, I have enjoyed talking about positive thinking, disability and DMD and sharing funny stories and my opinion. The friendly people at my Toastmasters clubs have shown me the benefits of genuinely sharing one's own experience. I have been going to Toastmasters for three years and it has enriched my social life. I have also learnt a lot from listening to my fellow Toastmasters.

Toastmasters is a level playing field where I can also educate, entertain and inspire my fellow club members. I am glad that people have found my speeches encouraging and interesting. My fellow Toastmasters and I learn from and enrich each other, and they have gained an understanding of some of the challenges I face in my day-to-day life. That includes needing help to breathe.

When I first started using a ventilator from the DHB more than ten years ago, it had a mask and I used it only at night. My muscles for breathing gradually got weaker, so I need to use it during the day also. Due to the humming noise generated by the ventilator, and the mask itself, it was difficult to hear and be heard clearly, especially with background noises. So I went to the Toastmasters

meetings without the ventilator, but I had to rush back to my van to use it again right after the meeting.

When I first started going to Toastmasters, I could be without the ventilator for three hours before headaches set in – I also had to endure a headache if I had a seven-minute speech to do. As I got exhausted from lack of oxygen, my breaths become shallower and my voice quieter. It really impacted my experience on every level.

Then the opportunity to use a more portable ventilator with a 'sip' attachment came, thanks to MDANZ and Resmed. I have been using the ASTRAL150 ventilator for nearly two years and it fits easily in a bag on the back of my power chair, with a tube that extends around to the side of my face. I can breathe from the 'sip' attachment regularly throughout the day.

Now I can go to Toastmasters, study, church, or visit my friends, and hear and speak clearly, so I don't have to constantly ask people to repeat their words, and vice versa. The sip ventilator, which makes much less noise, enabled me to continue my Toastmasters journey, and to speak at the 2016 Building Momentum and Disability Matters 2017 conferences.

I noticed the difference the sip ventilator makes when it was broken a few months ago. I had to use my old one with the mask, but unfortunately I could hear only half of what was said in the discussions in class. Thankfully, MDANZ got the sip ventilator repaired for me and I can again participate fully in what I love, such as Toastmasters and going to University.

I'm thankful for the support MDANZ has given me in on my journey. In 2013, when I was depressed, lonely and addicted to Farmville and currency trading, fieldworker Kristine introduced to me an inspirational book called *Life Without Limits* by Nick Vujicic. It led me to watch a video by the author that marked a turning in my life, where my addictions ended. She also encouraged me to connect with old friends and other MDANZ members, and to garner the support I need to achieve my goals.

Those goals included moving out of home and going flatting. When I couldn't find an accessible and affordable flat in Auckland in 2015, I moved to Hamilton. This was possible, because I had grown in confidence and courage through previous flatting experiences. When fieldworker Darian visited me in Hamilton, one of the things I talked to him about was my plan to study counselling. He too gave me the moral support and encouragement towards achieving whatever goal I really want to achieve.

Raymond has been to functions at many venues, including this one at Eden Park.

Now I can go to Toastmasters, study, church, or visit my friends, and hear and speak clearly.

My latest challenge is working towards a Bachelor of Counselling. I have taken on this new course of study after noticing how my fellow Toastmasters were encouraged by my speeches about experiences of having a positive attitude and coping with challenges. I feel that my personal experience of disability, depression and addiction will help me empathise with people facing these challenges. Also, being counselled has helped me learn about myself and improve my self-esteem. So as a Counsellor, I hope to help other people be happier and cope better in life.

Scenes from the conference. Photo credit: Cure SMA and Elaine Melko.

Community for a cure

Working together to make great things happen

In June this year, Fiona Tolich attended the Cure SMA Conference in Texas and discovered a wonderful community of action. She's come back determined to do everything she can to make a difference for New Zealanders living with the condition.

They say that everything is bigger in Texas and they are not wrong! Attending the Cure SMA Conference in Dallas had an enormous impact on me, and I strongly believe our community can work together to make big things happen.

I want to thank the wonderful people who pull this conference together – it really was phenomenal.

The impact it has had on my life is immeasurable. The conference sparked a fire I already had, now it's an inferno I hope never goes out. The feeling of community was immense, researchers, clinicians, families and individuals came together in a place where there was no judgement, just complete respect for one another, and understanding of what each other was going through.

Having travelled from New Zealand, Anna Sutherland and I covered the greatest distance to be there, but we were not the only ones. People from 30 different countries attended.

To help describe the magnitude of the conference, here are some other numbers:

- 450 researchers and healthcare professionals in attendance
- 55 family workshop sessions
- 185 researcher and care presentations

Taking into account that SMA is an 'orphan disease' (rare disease), it is amazing an event like this attracts so many people. This is a broad and large community, united in its mission to find a cure.

There were times I felt confronted with what this disease does to people (including being face-to-face with a child unable to breathe), but there were many moments of hope, hearing from those already on a treatment path, and those who have achieved so much with this condition. In many cases, beating the odds.

First up for me was the Newly Diagnosed session, where we were introduced to Kenneth Hobby, the President of Cure SMA.

Eighteen months ago, the community was all about hope, but treatment has helped that hope morph into practical reality with many in the room having access to Spinraza. This is what I want for our community at home – a chance to get our own piece of practical reality.

The exciting news is that more developments are in the pipeline and more treatments will be released. The thing about SMA is that we know the cause of it – missing the SMN1 gene. Other diseases have different versions and mutations, but we are the same (something that will undoubtedly help to make significant breakthroughs). It is the SMN2 gene that has given the drugs/treatments a target. There is some hope with researchers being able to manipulate this backup gene as everyone with SMA has it, it is just the copy numbers that are different.

The SMN2 gene makes a small amount of protein. If an individual has three or more copies of SMN2, research shows us that they are more mildly affected, if they have just 1-2 copies then they have enough to keep them alive through childbirth, but not enough to see them through life.

The mission for Cure SMA is a simple one; first slow the disease (current treatments are doing this), then stop it, and finally cure it. There is a lot of positive work going on, but there is so much more still to be done.

The key message from this session for me was don't wait. Don't wait for the next trial, or for a treatment easier to administer. The most important thing is what you can get now. New Zealand needs access to treatment, and alongside Anna Sutherland and our New Zealand SMA Reference Group, I am on a mission to drive it. Our people and our community are important, and we simply cannot be patient and wait!

We travelled with Julie Cini from SMA Australia. It is very exciting to see what she has achieved across the Tasman with access to Spinraza for those under 18. We wish them well with their next task of getting access for adults, and hope that in New Zealand, we can achieve the same things.

Photo credit: Cure SMA and Elaine Melko.

The conference sparked a fire I already had, now it's an inferno I hope never goes out. The feeling of community was immense.

Thomas Crawford, an SMA expert from John Hopkins Hospital in Maryland was another inspiring speaker. He described DNA as being like a string of pearls on stretches of string. DNA takes the necklace and makes a copy and the pearls have to be put together. The pearls represent exons. In SMA, one of those pearls is missing – exon 7.

He also explained that everyone has mutations, and 1/50 people have this mutation. Further to this, 1/2500 couples both have the same mutation. If you are unlucky, then SMN1 is missing from the child (think the pearl necklace).

The reality with SMA is that the weakness comes from the motor neurones, everything else is completely fine. The consequences of that is what we see with SMA – skeletal deformity, weakness, no strength to cough, respiratory difficulty and so much more. As more motor neurones die, the impact becomes greater on the individual.

Until there is a cure, SMA is described as being like the Mafia – once you are in, there is no getting out. I for one plan on getting us all out! A cure is possible and I believe it will happen in my lifetime! "Without faith, nothing is possible. With it, nothing is impossible", Mary McLeod Bethune.

The conference included the largest research meeting in the world specifically focused on SMA. The goal is to create open communication of early, unpublished scientific data, accelerating the pace of research. The meeting also furthers research by building productive collaborations—including cross-disciplinary dialogue, partnerships, integration of new researchers and drug companies, and educational opportunities for junior researchers.

Because the meeting was part of the conference, researchers had the chance to interact with, learn from, and educate families affected by SMA.

As representatives from New Zealand and Australia, Anna and I (alongside Julie Cini and Zoe Watson) chose to go to different sessions to get as much information as possible. We're committed to sharing this information with our community. It will include nutritional recommendations, yoga therapy, aquatic physical therapy, orthopaedic management, and much more.

The final day of conference included the announcement that next year's Annual SMA Conference will take place between June 28 and July 1 at Disneyland in Anaheim, California. I'm keen to be there. What I learned from this conference was well worth all of the effort and sacrifice, and the tears when I left my kids behind. If you can make it, I highly recommend it.

The work is just beginning in New Zealand. I want to treat it like a sprint relay race – the quicker we get out of the starting blocks, the better our chance of achieving something great. But we need to work together to get to the finish line - for the sake of our community.

Fiona and Anna (left) received funding from the MDANZ Members' Discretionary Fund to help them travel to the conference.

Muscular Dystrophy New Zealand

Help us raise money without spending a cent.

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Community Power is a new power company that wants to make a difference to the lives of New Zealanders.

To do so, it shares a portion of its profits with charities such as the Muscular Dystrophy Association of New Zealand (MDANZ).

By switching your power company to Community Power, you will stay on your current rates, or pay less – you won't pay any more for power. And by simply paying your power bill, Community Power will donate a portion of their profits to MDANZ and acknowledge you for this donation. It won't cost you a cent more and you won't have to dip into your own pockets.

This is the most powerful fundraising initiative we've ever launched. We don't need your money, we just need your power!

I've made the switch. Will you make it too?

Ronelle

Ronelle Baker
Chief Executive

Yes I will! How do I switch?

1. For a free, no obligation quote, email a copy of a recent electricity bill to: **support@communitypower.co.nz**
2. We will let you know how much your donation will be and if there are any savings for you.
3. Then just say "Yes" to start donating at no cost to you, we do the rest.
4. Your electricity supply will not be interrupted, we will take care of everything.
5. If you receive a network company dividend or rebate you will continue to receive this.
6. If you receive a prompt payment discount you will continue to receive this.
7. We will advise you on each monthly invoice of the amount you have donated and of your total donations made since joining us.
8. **Remember, your donations come from our profits, not your pocket. Thank you for helping.**

In partnership with COMMUNITY POWER *Power of Together*
www.communitypower.co.nz/md

Deflazacort shows benefits over prednisone

Increasing dystrophin production.

A recent publication in *Muscle & Nerve* compared the efficacy and safety of deflazacort and prednisone/prednisolone from participants in the placebo arm of the Ataluren Confirmatory Trial (ACT) Duchenne muscular dystrophy study.

Duchenne muscular dystrophy participants treated with deflazacort had notably less decline from baseline in six-minute walk distance over 48 weeks, than those treated with prednisone/prednisolone.

"This publication supports the benefit of deflazacort in slowing the progression of Duchenne compared to other corticosteroids," said Stuart W. Peltz, Ph.D. Chief Executive Officer of PTC Therapeutics, Inc. "The data indicates that deflazacort should be the standard of care for all patients with Duchenne. The availability of deflazacort, a treatment that has the potential to alter the natural history of Duchenne, supports the need for early diagnosis in patients with this disease."

PTC Therapeutics acquired the rights to Emflaza (deflazacort) from Marathon Pharmaceuticals in 2017. Ⓡ

FDA Orphan Drug Designation for Spinocerebellar Ataxia

Safety and efficacy data compelling.

IntraBio, a U.K. based biopharmaceutical company, has received an Orphan Drug Designation from the US Food and Drug Administration (FDA) for its lead compound (IB1000) for the treatment of spinocerebellar ataxias (SCA), of which there are over 40 known subtypes.

Intrabio's Senior-Vice President Taylor Fields told Ataxia UK "Although we have only investigated the clinical efficacy of IB1000 in a small number of SCA subtypes, we were pleased that the safety and efficacy data was so compelling that the compound was granted a single designation covering all SCAs."

In May, 2018, IntraBio filed an Orphan Medicinal Drug Application for SCA with the EMA, which they hope to receive between July and September 2018.

This designation will greatly support IntraBio's plan to conduct a multinational, multicenter, placebo controlled clinical study for the treatment of inherited cerebellar ataxias. Details of the study, including the protocols and inclusion/exclusion, will be made available shortly. Ⓡ

Early intervention for DMD

Results consistent for those with older children.

PTC Therapeutics has announced positive data from its Translarna Phase II Clinical Trial in children as young as two years with Duchenne muscular dystrophy caused by nonsense mutations (nmDMD).

An interim analysis demonstrated that at week 28, the safety and pharmacokinetic profile for Translarna in children aged two to five years is consistent with that for older children. Clinical benefits were also observed at 28 weeks with Translarna, with decreases versus baseline in the time to run/walk 10 meters, climb four stairs, and stand from lying face up (supine).

The data showed that treatment with Translarna resulted in improvements in timed function tests and the North Star Ambulatory Assessment from baseline at weeks 28 and 52, with mean changes showing as much as a 25 percent improvement after one year.

The data at 28 weeks formed the basis of the recent positive opinion from the Committee for Medicinal Products for Human Use (CHMP) of the European Medicines Agency (EMA) to expand the current indication of Translarna to include children from two to five years of age. The data was presented at the International Congress on Neuromuscular Diseases in Vienna.

Translarna is the only approved treatment to address the underlying cause of nmDMD and is currently licensed in Europe for ambulatory patients aged five years and older.

"We are excited to demonstrate that Translarna showed an improvement over one year of treatment in patients with nonsense mutation Duchenne as young as two years of age," stated Stuart W. Peltz, Ph.D., Chief Executive Officer of PTC Therapeutics, Inc. "Irreversible muscle damage starts before the age of five. Early intervention is critical to maintain muscle function and delay disease progression."

Disappointing outcome for DMD trial

After reviewing the top-line results of PhaseOut DMD, Summit Therapeutics has decided to discontinue the development of ezutromid.

In an announcement, the company wrote; "We recognise that this decision will be disappointing for the Duchenne community, but the data from PhaseOut DMD were clear that ezutromid, while well-tolerated, did not provide a benefit to patients with DMD.

"These results are not what we had hoped for, and certainly not what we had expected based on the encouraging interim results from PhaseOut DMD. But, they provide a clear answer to the important scientific question of ezutromid's effect in DMD. The results provided no evidence that ezutromid is having a meaningful effect on slowing DMD progression. We therefore cannot support further development of ezutromid.

"We plan to explore ways that the information gathered as part of PhaseOut DMD can be made available to support other research activities in DMD for the benefit of the entire community. We believe that the future of Duchenne research is bright. There are numerous clinical trials and research studies taking place in this field, and there is hope on the horizon for all those living with DMD."

Medicines for rare disorders

PHARMAC signals a change in approach

PHARMAC has recently released a revised set of policy settings for medicines for rare disorders. In June they began calling for funding applications from suppliers of medicines for rare disorders under these new policy settings and have established a dedicated subcommittee to consider the applications.

Overall this is a positive step, but some concerns have been expressed in the community about aspects of the policy.

The policy settings include three principles, which are based on the differences of medicines for the treatment of rare disorders. These differences address the current market challenges for these medicines, and enable (where appropriate) a different entry into the pharmaceutical funding process.

The principles are:

1. The medicine has been approved by Medsafe, or an approved international regulatory authority, for the identified indication.
2. The disorder is a clinically defined disorder affecting an identifiable and measurable patient population with a prevalence of less than 1:50,000 in New Zealand.
3. The medicine is only registered for the treatment of the rare disorder, or if it is registered for other disorders (or is part of phase three clinical trials for other disorders), the cumulative prevalence across all indications still meets principle 2.

PHARMAC will determine whether applications meet the principles. Those that do will then be considered through the process. Other applications that do not meet the principles can still be considered through the usual Schedule listing process.

The 1 in 50,000 threshold does have implications for some of the conditions covered by MDANZ. This represents a low number (approximately 90 people in NZ), which in other countries would be considered ultra rare. In Europe a rare condition is one that affects 1 in 2000 or less.

We are concerned this threshold will limit the treatments eligible to apply for funding under the rare disorders remit.

The Rare Disorders Subcommittee of the Pharmacology and Therapeutics Advisory Committee (PTAC) has been established with leading experts throughout Australasia who have a special interest in rare disorders, including neurologist James Cleland and clinical geneticist Dylan Mordaunt.

The committee will provide clinical input on funding applications PHARMAC receives for medicines for rare disorders. The Subcommittee includes two PTAC members, to maintain links with PHARMAC's primary clinical advisory committee.

We anticipate that these changes will lead to improved access to medicines for people with rare disorders. Ⓡ

What is Facioscapulohumeral Muscular Dystrophy?

Understanding one of the more common types of muscular dystrophy.

Facioscapulohumeral dystrophy (FSH), is the third most common of nine muscular dystrophy disorders, affecting approximately 1 in 20,000 individuals. The term Facioscapulohumeral uses three Latin words to describe the characteristic features of the disorder. Facio means face, scapula means shoulder blade, and humerous is Latin for the upper arm. Muscular dystrophy refers to muscle weakness and wasting. Thus, in FSH the muscles typically affected are those of the face, shoulder blade and upper arms.

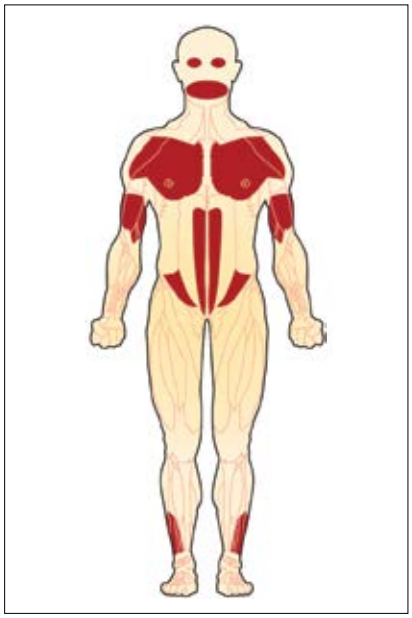
Features of FSH

FSH affects males and females equally. Symptoms of FSH can be classified into two groups: adult-onset, which is usually mild or congenital-onset, which is typically more severe and often present from birth. As well as variability of onset and severity, there is also variability in which areas of the body are affected. The classical symptoms of FSH involve the muscles of the face, shoulder-blade, and upper arms. Some people, however, have no symptoms in the face muscles, with the lower limbs affected instead. This is known as 'Atypical FSH'.

The following features may be displayed:

- Facial weakness, causing difficulty in pronouncing words, and using facial muscles – such as in whistling, smiling or closing one's eyes.
- Weakness in the shoulder blades, preventing movements such as throwing objects and raising one's arms. Lack of strength around the shoulder blades may allow it to wing out.
- Weak lower leg muscles, causing difficulties in walking up hills, or on uneven surfaces. Foot drop may occur when the muscles weaken to such an extent the front part of the foot cannot be lifted up during walking.
- Excessive spinal curve (lordosis), due to abdominal muscle weakness.
- Weakness in the muscles surrounding the hip and those of the upper leg, causing problems when rising from a chair, climbing stairs, or running.
- The development of contractures, as scar tissue replaces normal elastic tissue. This prevents normal movement in the joint, usually the ankles.
- Eye problems are infrequent, but more severe forms of FSH are associated with Coat's disease of the retina.
- Hearing difficulties are common; complete hearing loss may occur in severe FSH.
- Epilepsy in more severe FSH cases.
- Cardiac and respiratory problems are rare in FSH, but do occur in some patients.
- Inflammation of the muscles. This can be a source of pain in FSH, as can the altered joint position resulting from muscle weakness.
- Intellectual and cognitive (understanding) difficulties are very uncommon in adults with FSH. There are many FSH people, including those in whom symptoms began in childhood, in intellectually demanding occupations.
- Learning difficulties will on occasion occur, but it is important that real cases are not confused with apparent non-responsiveness. Very bright FSH children may appear intellectually challenged due to a combination of deafness and facial immobility.

FSH tends to progress slowly, and there may be long periods where relatively little change in symptoms



This diagram shows the muscle groups affected by FSH. Sourced from: http://mda.org/disease/fshmuscular-dystrophy/overview

FSH tends to progress slowly, and there may be long periods where relatively little change in symptoms occur.

occur. It may take 30 years for serious problems to develop, if at all. Up to 20 percent of people may require a wheelchair for mobility. Life expectancy is that of the general population.

What causes FSH?

The cause of FSHD is complex. There are two main causes of facioscapulohumeral muscular dystrophy (FSHD) resulting in either FSHD type 1 or type 2. The most common is FSHD type 1 which accounts for 95 percent of cases. Although FSHD1 and FSHD2 have different processes they both result in the same end outcome, which is the switching on of the DUX4 gene. This gene is usually switched off in muscles because once activated it causes muscles to be broken down.

FSHD1

Facioscapulohumeral muscular dystrophy type 1 is mostly inherited as an autosomal dominant trait. In most cases, the affected chromosome is inherited from a parent, but sometimes the variation occurs spontaneously during early development in the womb. The genetic change affecting people with FSHD1 is located on chromosome 4 where there is normally between 11 and 110 repeat units of a genetic motif called D4Z4. In people with FSHD1 the number of repeat units is only between 1 and 10, a 'repeat contraction'. The D4Z4 units are very important because they act as a barricade, preventing DUX4 from being switched on. When there are fewer DZ4Z units, the DUX4 gene is allowed to be switched on.

FSHD2

Facioscapulohumeral muscular dystrophy type 2 is called a di-genic disorder because it requires two 'hits' or mutations in your genome. One of these mutations needs to occur in a gene called Structural Maintenance of Chromosomes Hinge Domain Containing 1 (SMCHD1) on chromosome 18 that results in reduction of the protein it produces. The SMCHD1 protein plays an important role in locking out regions of your genome, one of these regions contains DUX4. Reduced SMCHD1 protein means that not all of DUX4 can be withheld and the gene is able to be switched on.

Before DUX4 can be switched on though it requires a particular genetic sequence – a second mutation - to enable the conversion of the DUX4 gene into the destructive DUX4 protein. So, second mutation next door to DUX4, known as a pLAM sequence makes it "permissive". FHSD1 is autosomal dominant because only one copy of chromosome 4 containing both the repeat contraction and a "permissive" pLAM is required and this can be passed on from a single parent. A person with FSHD2 can inherit the mutations separately; the SMCHD1 mutation on chromosome 18 from one parent and the "permissive" pLAM on chromosome 4 from the other parent.

FSH is an autosomal dominant disorder in as many as 70-90 percent of cases, meaning only one copy of the defect is required for the FSH to develop. This is in contrast to an autosomal recessive disorder, where two copies of the defect are required for the disease to develop.

The remaining 10-30 percent of defects arise spontaneously, with the deletion occurring by chance in the egg or sperm at conception. With a spontaneous mutation, the affected person will be the first in his family to have FSH.

There is a 50 percent chance the child of a FSH parent will inherit the mutation and therefore develop the

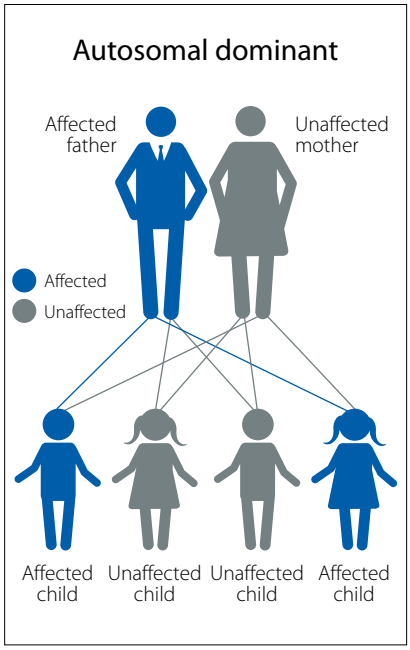
condition. Females are affected just as frequently as males, although symptoms in men are generally more severe and occur at a younger age than in women.

For further information on genetics and how disorders are inherited, please refer to the Muscular Dystrophy Association Genetics Fact Sheet.

Diagnosis of FSH

There are often difficulties in diagnosing FSH, as symptoms vary. However, once FSH is suspected, diagnostic tests will be offered to establish a definite diagnosis. These may include:

- **DNA Testing**
  There is now a reliable DNA test for FSH, which is approximately 98 percent accurate as a presumptive diagnosis. Laboratory technicians are able to extract DNA from a small amount of blood, and detect the DNA deletion responsible for the disorder. DNA testing can be done during pregnancy to determine if the fetus has inherited the deletion for FSH, but the deletion size can not be used to predict accurately the severity of the condition. The testing can raise ethical issues for parents faced with the option to terminate a pregnancy.
- **CK Testing**
  A blood test can assess the presence of an enzyme creatine kinase, also known as creatine phosphokinase. This enzyme is usually restricted to muscles cells, but when muscles are damaged as in FSH, the enzyme leaks out and into the blood serum. The CK test will show elevated amounts in the blood, but is inconclusive as elevated CK is also a feature of other forms of muscular dystrophies.
- **Electromyogram (EMG)**
  An EMG measures the electrical activity of muscles, and also measures the muscle's response to stimulation of its nerve supply. The results may be nonspecific, or show both nerve and muscle involvement, which is typical of FSH.
- **Muscle Biopsy**
  While under local anaesthetic, a small amount of muscle tissue is taken with a needle, usually from the thigh. Using special staining techniques in the laboratory, the muscle tissue is examined microscopically. This can give a lot of information on the condition of the muscle, and can help to rule out other diagnoses, or confirm the FSH diagnosis.



Soon after a diagnosis of FSH in the family, it is essential that genetic counselling is arranged.

Other tests include nerve conduction velocity, hearing tests and tests of cardiac function.

Soon after a diagnosis of FSH in the family, it is essential that genetic counselling is arranged, for one or both of two issues. The first is the probablility of Mum or Dad having the disorder, and the second is whether testing for FSH in pregnancy can be offered and with what degree of accuracy. Genetic counselling provides information about possible diagnostic tests, including prenatal testing. Genetic services in New Zealand are available and a referral can be made by the MDANZ.

Management of FSH

As yet, there is no known cure that can halt or reverse the symptoms and progressive muscle weakness associated with FSH. It is possible, however, to control complications by adhering to a management programme specially designed by a team of medical professionals. This team may include occupational therapists, physicians, orthopaedic surgeons, physical therapists, orthotists, dietitians, nurses and psychologists. Many other people are

there to give advice and help in any way possible, such as social workers, teachers, religious advisers, staff from the MDANZ, parents, and other persons with FSH.

- **Exercise**
  Moderate exercise, especially swimming, is generally considered to be beneficial in FSH, maintaining both muscle strength and flexibility, without undue strain. Swimming is a great source of active exercise as the water can be used for support. Passive exercise, or assisted stretching, should be established as early as possible. Physiotherapists will be able to assist in the development of an exercise programme to delay the shortening of muscles (contractures), which causes limitations in the range of motion of joints. These exercises should be undertaken on a daily basis and require assistance from parents and/ or caregivers.
- **Supportive equipment**
  Braces and splints are likely to be required to help compensate for weakened muscles. These are often worn at night to help maintain joints in a normal position. Other types of supportive equipment will be available as the need arises, and usually caters to individual need. Advice concerning these will be offered by the physiotherapist, occupational therapist, or by the MDANZ.
- **Medical treatment**
  Drugs such as albuterol, clenbuterol, and oxandrolone are being studied for their muscle building effects. These treatments seem to be moreeffective in the early stages of FSH, improving some measures of strength. Anti-inflammatory drugs may be prescribed to reduce associated inflammation.
- **Nutrition**
  Excessive weight gain leading to obesity can occur due to reduced physical activity produced by muscle weakness. Any excess weight will contribute to tiredness and weakness; hence it is important to maintain a good balanced diet incorporating plenty of fresh fruit and vegetables.
- **Surgery**
  Stabilising the shoulder blades is one of the more common surgical procedures undertaken by FSH patients. The winged scapulae are fixed to the ribs, so they don't move around. Although the surgery may actually decrease the arm's range of motion (since the shoulder blade can no longer rotate normally), the ability of the arm to function may be better, as the arm's leverage point is now stable. Tendons can be surgically severed to relieve contractures. This operation is most often performed at the ankle joint, but will also benefit those that have already developed severe contractures in the knees and hips. Surgery on the eyelids may be beneficial where there is incomplete closure of the lids. Incomplete closure of the lids may cause inflammation of the cornea (keratitis), so it is important not to ignore the early signs of waking up with dry and irritated eyes. Surgery may produce significant benefits, although these must be balanced against potential complications. Postoperative immobilisation can cause further muscle wasting, and extensive physiotherapy will be required after some surgeries. ®

It is important to maintain a good balanced diet incorporating plenty of fresh fruit and vegetables.

Finding my smile

Lisa Howe shares her journey with FSHD, what she's learnt from the members of her family, and what she's discovered for herself.

My dad always knew I had FSHD. As a baby, I'd sleep with one eye half open, only ever drooling on one side of the pillow. I never thought too much of it, seeing the condition more as an interesting familial trait than anything I should worry about. My dad and grandad are affected, along with two aunties who are part of the MDANZ team (Hi Aunty Ronelle and Tonya!), so FSHD has never felt particularly scary to me. Having type 1 diabetes has also made me a bit more brave, what with all the needle injections and the blood tests. It wasn't until I was 14 and a nurse asked me what other conditions I had, that I realised I'd never actually been diagnosed with muscular dystrophy.

After that, I saw a neurologist who did all the official tests. I felt oddly comforted when my results came back, affirming what my Dad and I had always believed. However, the condition had changed for me, becoming something medical and serious and definitely scary. I realised a lot of things I couldn't do that I hadn't really noticed before, or that I had just attributed to my nerdy, non-athletic self. I couldn't whistle, couldn't blow up my cheeks, and had difficulty raising my arms above my shoulders. My twin sister was always the sporty one, and I'd just assumed everyone had trouble washing their hair sometimes. There was also my crooked smile, which the doctor quickly pointed out as a classic sign of FSHD. I began to notice that smile in every photo, and in every video of me laughing. There aren't many pictures now of me smiling in high school.

I remember doctors telling me about IVF options in the future, in case I was worried about my children having the same condition. At the time it seemed funny, I was barely thinking about boys, let alone having babies. I'm still far off now, aged 20, but I do wonder what my future will look like. The condition is (thankfully) progressing quite slowly, but I do try to maintain strength through regular exercise. My current boyfriend is very active, so that encourages me to keep up with him! He knows about my muscle weakness and it's never really been an issue, but I do think about how things might change. I'd love to be a mum someday, with a supportive partner who understands my condition, and can balance out any physical difficulties.

I've always been interested in the mind, and how much there is to learn. Sharing knowledge is a passion of mine which I would love to harness one day as a teacher. I am in my third year at the University of Auckland, studying a conjoint Arts/Science degree in English and Psychology. While I appreciate what my body is able to do for me, I don't believe that it defines who I am. I have many lovely family members with FSHD and I don't feel that the condition has negatively affected who they are as people. Personally, it's allowed me to find value beyond my physical appearance, focusing instead on positive characteristics that radiate outwards (as cliché as that may sound).

Lisa (right), with friend Hannah.

In my last year at high school, I found a video online about FSHD. The woman in it shares the difficulties of her condition and, most importantly, how hard it was for her to lose her smile. Though it's still rare for me to bear my full-fledged grin on camera, I have no trouble laughing wholeheartedly in front of other people. No longer does it feel like an unflattering sign of weakness. Instead, all I am is a happy girl with her smile.

As Roald Dahl says; "A person who has good thoughts cannot ever be ugly. If you have good thoughts they will shine out of your face like sunbeams and you will always look lovely."

Support boosts independence

LIZ CHURCH

It turns out that a little help in the morning, can mean a more productive day. Liz Church shares her invidualised funding story.

It was great to catch up with Ronelle Baker recently at a work-related dinner. We last met about three years ago at the MDANZ conference. Ronelle had just been employed as National Service Leader, but had not yet begun her new role. I was attending the conference independently, with just my stick for support. This time Ronelle was the head honcho (or Chief Executive to give her official title!) and I was using a wheelchair, and employing support staff. It really brought home to us both how quickly our circumstances can change!

Back then, my trusty stick and I could go most places alone as long as they were accessible. So what's changed? Well, I do have a slowly progressive condition but the turning point was a random fall I had in January 2017. After surgery for a broken hip, I had to learn to walk again. Ironically it was quite a positive experience. While my mobility had been on a downward trajectory for decades, here I was getting more mobile as the weeks went by. Go figure! By August I had said goodbye to my physio, in November I reached a high point doing a little Christmas shopping in town, just me and my stick again. It was short-lived success, but enjoyable nonetheless.

I always thought you had to have a Community Services Card to be eligible, and that working meant I didn't qualify.

Another fall in January 2018 didn't injure me but it stole my confidence, and shrank my world again. It'll be no surprise to you that it's harder to make a case for support when there are no x-rays to back you up! I did get physio support in the end, but my big 'aha' moment came when I discovered I could get some personal care support. I always thought you had to have a Community Services Card to be eligible, and that working meant I didn't qualify. I felt a bit silly not knowing that - because I work in the disability sector (though not in services). It was a classic case of 'you don't know what you don't know'.

So now I have personal care support through individualised funding [IF] which means I have an annual budget, rather than allocated hours. This support sets me up so I can have a more productive day. It was also easy-peasy to arrange extra support on that trip to Auckland when I caught up with Ronelle. It's a bit overwhelming learning my responsibilities as an employer but I have the support of a coach, helpdesk and lots of resources. I've also found online forums where people using IF share their collective knowledge. If one person doesn't know the answer to your question, someone else will. It's a great reminder that having independence doesn't always mean doing everything alone.

Liz Church is originally from Whanganui and has had over 25 years' experience in early childhood education. She has worked in the disability sector for ten years in various roles, currently employed as a project coordinator.

Am I free?

SCOTT BOYLE

Sometimes, a rebellious streak can lead to positive outcomes.

Looking back on my childhood and teenage years I realise how much who I am today has been shaped by my own personal liberties. My relationships, decisions, successes, and mistakes were all deeply affected by my ability to live the way I wanted to live. It was clear to me early on that the secret to living a long life with my condition was being able to simply enjoy that life. So I fell in love, partied, socialised, protested, and travelled. I spent years soaking up all I could in the world because I knew inevitably the day would come when my condition would begin to take a toll and wear me down. A lot of people aren't as lucky though. Their situation restricts their freedom at a young age, and that infuriates me. So I've come to appreciate my own freedom a great deal more, and intend to use it wisely.

When I was 17 and in my last year of high school, my closest friend, who is like a brother to me still, asked for my help. Our school had decided not to do its annual theatre production and as theatre students, we were outraged by the decision. Our final chance to perform in a show with dozens of our friends was gone. How could we be robbed of the thing we were most looking forward to in our senior year? There was a dark cloud over the drama department from that moment on, and few of us felt enthusiastic anymore when we attended classes. So we decided to produce our own show.

Two teenagers with no experience in producing theatre shows chose to do what nobody else was willing to do. It was an act of defiance that sent a ripple throughout the school. For us, it wasn't about going against the school or proving a point. It was about exercising our freedom to challenge ourselves and provide a memorable show for our friends. We wrote a script, put together a cast, acquired props and costumes, and spent months rehearsing and directing in preparation for the big day.

Our initiative was enough to reinvigorate every drama student, from Year 9 to 13. The department had been a somber and desolate place for a fortnight but now, it had returned to life. The most vivid memory I have of that show was spending two days trying to convince our principal to let us use the school drama wing for rehearsals and performances. The teachers had little faith in us. But as I learnt, being underestimated is an incredible gift. Somehow we managed to put on three shows, followed by an encore night, and raised more than $5000 for the Make A Wish Foundation. When the adults didn't want to attempt a show, we made the choice to accept the responsibility and make it happen, creating change in the process.

Looking back, I recognise my determination to do something memorable was mostly thanks to my parents. From the moment I was diagnosed, they made the choice to empower me and let me make my own choices. While some wanted to treat me as a fragile porcelain doll, they treated me like any other kid. They taught me to have my own mind and not be like everybody else. That guidance encouraged me to figure out what my passions were, and choose a path I wanted instead of following others down a road I cared nothing about. My parents instilled me with an understanding of freedom early on, and gave me the tools to figure out who I was. While at times I may stumble or make mistakes, I am grateful for the freedom to experience such failings and find a way forward.

The teachers had little faith in us. But as I learnt, being underestimated is an incredible gift.

Scott Boyle lives in Christchurch and is the Rangatahi Representative on National Council. He loves history and storytelling, and is currently working on his first novel. He is passionate about raising awareness of muscular dystrophy and ensuring younger voices are heard.

Ask the **PANEL**

Treatable neuromuscular conditions

DR. RICHARD ROXBURGH

What do we mean by the word treatable?

We could mean treating the symptoms of a condition or managing the condition to make the most of what someone has, or we could mean treating the cause and preventing the condition altogether or reversing the effects of the condition.

Neuromuscular disorders are acquired or genetic conditions that affect some part of the neuromuscular system. Neuromuscular disorders tend to be progressive in nature, and result in muscle weakness and fatigue. Some are present at birth, some manifest in childhood, and others in adulthood. It may be genetically passed down (inherited) or due to a spontaneous genetic mutation, or may be due to an abnormal immune response, inflammation, poisoning, toxins or tumours. Some disorders have no known cause.

Neuromuscular disorders can be categorised according to what part of the neuromuscular system is affected; nerve cells in the spinal cord (motor neuron diseases), nerve fibres running to the face, neck, arms and legs (peripheral nerve diseases), the site where nerves and muscles meet, and diseases which affect the muscles themselves.

Disorders are also classified by the disease mechanism. For example, muscle disease could be caused by

- Inflammation (e.g. dermatomyositis)
- Degeneration (e.g. inclusion body myopathy)
- Metabolic dysfunction (e.g. McArdle's disease where the body lacks an enzyme to make glucose)
- Genetic, where one of the proteins for a muscle's integrity or structure is lacking (e.g Duchenne muscular dystrophy) or not working properly (e.g. Becker's muscular dystrophy)

Treatment options are different depending on how the neuromuscular system is affected. An abnormal immune response, for example when a patient's own immune system is attacking healthy muscle tissue, is usually treatable with medications that suppress the immune system. Inflammation can usually be treated with anti-inflammatory medications.

The processes that are in play in genetic conditions involve loss of protein (loss of function) or toxic gain of function of a protein. These conditions are often progressive; getting worse over time. Treatment may just slow or halt the process - but won't necessarily be able to instigate regeneration of the damaged nerve or muscle.

In some genetic conditions replacement of the missing protein, for example replacement of the enzyme missing in Pompe disease, works to alleviate the progression of the disorder.

In other genetic conditions pinpointing the exact cause of the disorder has also led to knowledge of how the disorder can be treated.

The best current example we have of this is a remarkable new treatment now developed for spinal muscular atrophy (SMA). SMA occurs when a person has an altered survival motor neuron type 1 (SMN1) gene that means they are missing an important protein called survival motor neuron protein. SMA affects the anterior horn cell and once degenerated these cells are no longer able to innervate the muscle and so the muscle eventually wastes away (atrophies). There is a second gene though that makes very small amounts of working survival motor neuron protein, called SMN2. The new treatment tricks SMN2 into forming more of the SMN1 type protein.

A number of other genetic conditions have similar kinds of treatment currently being developed and trialled, including Duchenne muscular dystrophy, myotonic dystrophy and Huntington's disease.

Accurate genetic diagnosis is crucial to being able to receive such types of treatment.

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.

Caution- allergy season ahead!

MIRIAM HANNA

Tis the season to sniff and sneeze

With spring just around the corner, many of us start to get anxious about allergies that flare up during this season. By many, I mean a whopping third of us. New Zealand has one of the highest prevalence rates of allergies in the world. The underlying cause of allergies is unknown, however there are many theories including: our environment and diet in the first 1000 days of life from conception; family history and genetics; our diet and the effect it has on our gut microbiota, the effect that has on our health; exposure to sunlight and Vitamin D levels; as well as exposure or lack of exposure to bacteria in the environment.

The latter theory is commonly known as the "hygiene" theory. Scientists have discovered that a higher exposure to bacteria in our environment is correlated with lower rates of allergy and asthma. People from rural areas, including farms with arguably a higher exposure to bacteria, tend to have much lower levels of allergy and asthma than those who grow up in cities. In particular, those who live in very hygiene-conscious environments, where sanitisers and disinfectants cover every surface, have a particularly higher level.

This column is focused on environmental allergies or allergens, rather than food allergies. An allergen is a usually harmless substance that is incorrectly recognised as harmful in a person with allergies, resulting in the immune system triggering a response leading to an allergic reaction. Common environmental allergens resulting in hayfever (rhinitis) and asthma include pollen, dust mites, animal dander and mould.

For many of us, symptoms of hayfever and asthma can be debilitating with a huge effect on daily life. The best way to minimise the symptoms is avoiding the allergens or triggers if they are known to you, although this isn't always easy. First, find out what triggers or worsens your hayfever and/or asthma by asking your GP to request a skin prick test, performed at certain Labtest branches. This very straight forward procedure isn't painful, and tests for the most common allergens. Once you know your triggers, you can work out how to minimise them.

If you are allergic to house-dust mites which are commonly found in bedding or furnishings, it might be worthwhile investing in allergen-barrier sheets and pillow covers, now sold at a number of retailers. If animal dander is a trigger, it might be best not to have pets around your home, and ask if others have pets before visiting. For more information, and tips and advice on how to avoid your allergy triggers or minimize contact, you can contact Allergy NZ or visit www.allergy.org.nz

Allergy symptoms are often controlled with medication. For symptomatic relief of hayfever, your GP can prescribe antihistamines. Many antihistamines are also available without prescription from your local pharmacy. If you need to drive after taking antihistamines, it is important you take a non-sedating one, meaning it will not make you drowsy. It is also a good idea to discuss a steroid based nasal spray with your GP. Sprays are recommended as ongoing daily therapy to control your body's reaction to the allergens (as opposed to antihistamines which offer instant symptomatic relief). Flixonase is a common brand of nasal spray. It is important to start well before the allergy season, and use daily - even when you don't have symptoms - to achieve the best control.

If you use an asthma inhaler, talk to your GP about your dosage and whether you are using the best one for you.

Miriam Hanna has recently left her role as Member Services Manager at MDANZ to return to work in the pharmaceutical industry.

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

Melanie Kerr
Executive Assistant

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

Wellington Branch


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

Canterbury Branch


Fieldworkers: *Paul Graham and Marty Price*
Office Manager: *Gemma Foulds*
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

Wellington 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

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