National and international news and views
Networking Dystonia patients

New Zealand Dystonia Patient Network Inc.

News+Views

Newsletter 96 – August 2023 • Contact us: 0508 397866 or info@dystonia.org.nz • Donations are welcome

Message from the Chair – Alison Fitzpatrick

Alison is currently overseas enjoying a well deserved break so we take this opportunity to thank her for all she does for your Network, taking on Website management and other roles and steering us in the right direction.

One of Alison's many skills is opening and running our Seminars, deftly introducing and attending to our speakers, often with well-timed humour making everyone, including our speakers, at ease. This seminar was no exception.

Safe travels back home Alison, you can have your say in our next newsletter!

2023 Seminar and AGM

We had yet another successful seminar on 17 June at the Sudima hotel, next to Christchurch airport, in fact we were so engaged with all our speakers that we forgot to take any photos, our apologies!

We would like to take this opportunity to thank all of our speakers for donating their valuable time to provide the attendees with invaluable and easily understood information.

We provide the following summaries of our speaker's addresses:

Dr Tim Anderson

We were treated to a very interesting address by Dr Anderson and delivered in such a manner that made it easy to understand, particularly in the area of genetics.

Dr Anderson started by explaining some types of movement disorders: Tremors, Jerks and Spasms. It is Dystonia that is aligned to spasms he explained and described as:

- Sustained or intermittent muscle contractions causing abnormal, often repetitive movements, postures or both
- Dystonic movements are typically patterned, twisting and repetitive
- Dystonia is often initiated or worsened by voluntary action and associated with muscle overflow activation

He noted that dystonia has been re-classified into what is known as *Axis I* and *Axis II,* comprehensive listings of the different types of dystonia and what the cause may be.

Axis I covers Clinical Characteristics – such as age at onset, body distribution, temporal pattern and other neurological or systemic manifestations.

Axis II, covers Aetiology (the cause or origin of dystonia) – such as looking into the nervous system pathology (degeneration, structural lesions) or it may be inherited or acquired through brain injury, infection or drugs etc.

Dr Anderson explained the various types of dystonia (body distribution) with examples (ensuring anonymity), some before and after botox showing definite improvement. Interesting points raised were:

- Children presenting with focal dystonia have a higher tendency to progress to generalised dystonia. Individuals with a late onset will commonly remain stable in their dystonic presentation.
- Dystonia of the extremities is more likely to have a genetic origin
- Women are overall more likely to develop dystonic signs than men

Tim also talked about genetic dystonia and gave a very interesting insight into identifying particular dystonia genes within families and relatives.

Currently, samples are sent overseas for classification and a lot of progress has been made in this area.

One interesting question from a member put to Dr Anderson was "*is cramp linked to dystonia*". This was not considered to be linked unless the cramp was of a spasmodic nature, for instance recurring and not a temporary discomfort.

He concluded by noting that dystonia is the second most common disorder he sees, Parkinson's disease being the most common.

We thank Dr Anderson for taking time out from his no doubt busy schedule to address our membership with a very enlightening talk.

Dr Jacqui Allen

We enjoyed a lively and informative presentation by Dr Jacqui Allen. She is a laryngologist both in private practice and in the public system, where she runs a clinic for voice patients at two facilities, North Shore and Waitemata Hospitals. Dr Jacqui explained that although she is in the ENT field, she specialises in the 'T' of 'ENT' - that is, the throat. Her presentation covered dystonias that affect the neck, shoulders, throat and tongue. She made the point that severe cervical dystonia can cause problems with eating.

Dr Jacqui said that the vocal cords have two main, and competing, tasks – one is to allow breathing to take place, which involves opening the cords, and the other is to close off the cords, so that food and liquid avoid the airway to the lungs. Their function in vocalising is very much an after-thought, but of course extremely important for human communication.

When a patient presents to a laryngologist the throat area is examined internally, using endoscopy with cameras and tubes. In dystonia the vocal cords look normal, so the diagnosis relies on the doctor listening carefully to the voice - looking out for the straining and breaks typical of laryngeal dystonia (LD). There is no blood test for dystonia, or anything that looks unusual in a scan, so it is difficult to diagnose.

Dystonia was originally considered to be a disease of the basal ganglia but there is now a revised understanding of the process. There is a loop of activity – within the brain – where the signaling to the target muscles (vocal cords in LD) is not happening correctly. In particular the inhibition of various movements is abnormal.

Because the problem is in the brain, treatments such as botulinum toxin only control the symptoms.

Using sensory tricks, or gestes, are a common way of relieving symptoms. It is thought that these distract the faulty loop in the brain, hence showing that central processing in the brain is indeed the problem.

Dr Allen believes that advances are most likely in the field of genetics, and that perhaps gene editing will be possible in the future. Genetic research shows that only 12% of patients have a family history of dystonia.

Dystonia often co-exists with other disorders, such as essential tremor.

Jacqui talked about botulinum toxin. There is huge variability between patients – with all the factors of site, dose and duration. It takes about 2 days to shut down the release of acetylcholine from the muscle receptors. These then take about three months to regenerate - hence the typical timeframe of benefit from an injection. The botulinum toxin induces a flaccid paralysis of the vocal cords, making them floppy. If the paralysis was rigid, this treatment would not work. There are three main manufacturers: Allergan, (who make the trade-marked *Botox*®), *Xeomin*, and *Dysport*.

There is a promising new drug treatment on the horizon, although it is not yet available in New Zealand. It is now used 'off-label' in the US, following successful clinical trial. It is sodium oxybate, with commercial name Xyrem. This drug has the benefit to control LD, and could be used either on its own, or in conjunction with botulinum toxin to extend the effect. It seems more useful for patients who experience some benefit from moderate consumption of alcohol. Other treatments in future could include vibrotactile stimulation - delivered through a collar worn around the neck. Trials have shown that if this buzzing sensation is applied for 15 minutes then the voice improves for an hour or two.

The Network appreciates Dr Allen's willingness to travel down from Auckland to Christchurch and back on a Saturday in the depth of winter and deliver such an engaging and insightful talk to us all. She is willing to answer questions at jeallen@voiceandswallow.co.nz.

Laraine McAnally

It was a pleasure to have one of our Aussie cuzzies from the DNA at our seminar. This is the first time since 2019, so welcome back cobber!

Laraine began by thanking the NZDPN for inviting her to the seminar, stating how much the DNA values our alliance. She then went on to outline new initiatives the DNA has put in place over the last couple of years. As with all of us the way we get our information and stay in contact with folk has changed due to the pandemic. Some things that the DNA are doing differently include;

- Using zoom for online communication
- Creating a DNA YouTube channel
- Making the website more user-friendly

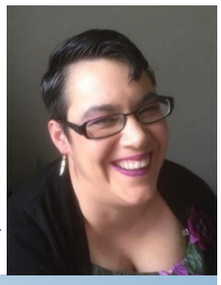
Another project was the award of three research grants to support trials for Dystonia treatments. The grants were made possible by people who fundraised for the DNA. Then the DNA rounded the sum up to $5,000 dollar amounts for each. With trials in TMS: Transcranial Magnetic Stimulation and MRgFUS (note the article in this newsletter on MRgFUS): MRI guided focused ultrasound technology; it is hoped that in the future dystonians will have more treatment choices.

For more information visit www. dystonia.org.au

Anna Nelson

We have had the privilege of Anna addressing us at our last seminar and time certainly hasn't dampened her spirits or enthusiasm!

Anna has a very much can do attitude and does not let her Cerebral Palsy get in the way of what she sets out to achieve, such as applying for her managerial role at AUT University, in which she was successful - Practice Manager, Disability Support (Anna is now the Service Manager – Disability Support Service) – and obtaining her driving licence.



Anna again talked without notes and gave a very heartfelt and emotional story of her experiences, the room was noticeably quiet when Anna spoke of her personal challenges, we all wholeheartedly supported her and praised her attitude.

Anna's Techniques for Keeping Positive and Navigating Barriers.

- Reach out to others.
- Remind myself how far I've come, and how much I've achieved, like learning to drive.
- Socialise as much as possible.
- See the humour in situations.
- Enjoy coffee and wine.
- Be in tune with what works – do I need time out, or time in the sun?
- Remember that others have confidence in me.
- Be mindful of limiting language like "handicapped."

And finally, Anna's motto:

Life can be cool if we make it cool.

Elisabeth Wells

Elisabeth generously gave us her time to address us on aspects of Laryngeal Dystonia (formerly known as Spasmodic Dysphonia).

Although Elisabeth was not on our scheduled list of speakers at the time of print of our newsletter she contacted us wondering if she would be able to do a brief talk, which of course we agreed to and just as well we did as she delivered a well thought out PowerPoint presentation.

Thank you Elisabeth for stepping up!

St Vincent's commences Australian-first trial for the treatment of focal dystonia

28 Apr 2023

Using state-of-the-art MRI guided focused ultrasound technology, St Vincent's Neurologist and Movement Disorders Fellow, Dr Joel Maamary is leading a clinical trial investigating the safety and efficacy of MRI guided focused ultrasound for the treatment of focal dystonia.

Focal dystonia a type of movement disorder, characterised by sustained or intermittent muscle contractions resulting in abnormal movements and/or postures that are isolated to a single body part. This type of dystonia is commonly triggered by particular tasks, such as writing, or playing an instrument. In these conditions, abnormal signals are generated within the brain, generating involuntary muscle contractions and an inability to perform the task.

With limited treatment options offering low or temporary relief, individuals are often left restricted and unable to perform tasks associated with their careers and passions. Purchased in 2018, the St Vincent's MRgFUS, the first of its kind in the Southern Hemisphere, has been used highly successfully for the treatment of movement disorders such as Parkinson's disease and essential tremor, and has now been extended to trial in the treatment of focal dystonia, with the hope of providing permanent relief for sufferers.

On 26th April, Ella Laskova, who has been living with hand dystonia for some years, became the first musician to undergo this treatment in Australia. Learning to play the violin at age 7, Ella became a celebrated chamber musician performing across Europe, but the impact of her condition left her without full movement in her left hand and unable to continue at an elite level. Unfortunately, her condition went unrecognised and she endured several operations and treatments on her left arm, without any improvement.

Using MRgFUS, the team created a small lesion to disrupt the abnormal brain pathways driving her dystonia. Ella experienced immediate improvement in her left hand movements following the procedure, with no side effects. "Ella's successful treatment demonstrates the power of this novel technology and provides hope for countless individuals who struggle with this condition on a daily basis." Said Dr Maamary.

The trial involves an in-depth analysis of the changes following treatment, essentially the rewiring within the brain to alleviate this debilitating condition, as well describing the cognitive changes noted within the brain. Additionally, the team is performing a rigorous analysis of upper limb strength, coordination and function to help better quantify both the effectiveness and side effect profile of this treatment. It is hoped that the information gained will assist in informing the scientific community to help transform the lives of individuals living with dystonia.

For the full article and link to the video: https://www.svhs.org.au/media/news/st-vincent-s-commences-australian-first-trial-for-the-treatment-of-focal-dystonia

(We have just heard that a patient has had successful surgery with this procedure, hopefully an update on this in our next newsletter).

David Barton

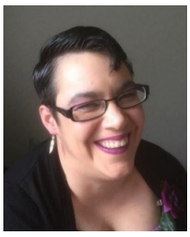
We reluctantly bid farewell to David from our Committee to a well-deserved retirement.

David has been on the Committee at least four times since its inception (David was instrumental in this) some 30 years ago and when not on the Committee has provided invaluable support as Database Manager, crafting letters and documents and latterly as our Treasurer. It is an understatement to say we will miss you David but hope to catch up from time to time.

On behalf of our Committee and our membership we send our best wishes.

New Committee Members

We thoroughly welcome Anna Nelson, Julie Hooke and Kylie Miller to our Committee, we had been looking for new members for some time so we are delighted they have made themselves available, welcome aboard!



Anna Nelson

Anna doesn't really need any introduction, she has become somewhat of a regular in our Newsletter!

And see the summary of her address at the seminar.

Kylie Miller

My journey with dystonia is relatively new. I noticed a head tremor and neck pain around a year and a half ago and was diagnosed with cervical dystonia in February this year. The diagnosis has definitely reduced my fear and given me greater clarity. Botox treatment has been really effective for me so far. It was so helpful to meet others with dystonia at the June Christchurch meeting. The committee are a lovely, supportive and fun group. It is so beneficial to have access to the avenues for support and information that the network provides.

Julie Hooke

My journey with Cervical Dystonia started roughly 6 years ago but when I look back it probably started in my 30's! Pain and stiffness in the neck which graduated to a tremor. Once diagnosed I tried medication but the side effects were dreadful. Tried massage and other alternative therapy which really didn't help me. I haven't tried botox but I do take anxiety medication, which is helpful to relax my muscles especially with everyday work stresses. Meeting everyone at the Seminar recently in Christchurch was just what I needed. Such an informative day with the bonus of meeting new friends who understand what you are going through or have gone through. Highly recommend attending if there is one in your area.

Governance of the NZDPN

The Executive Committee is elected each year at the Annual General Meeting. The Committee for 2023 is

Chair: Alison Fitzpatrick
Secretary: Desiree Sargon
Treasurer: TBC
Committee Members: Jayne Lewington Lovell, Dave Mitchell, Kylie Miller, Julie Hooke, Anna Nelson

The network is always in need of members to assist in dystonia projects. To keep the group running and to support other dystonians, please come and join us. Contacts are 0508 397 866 or info@dystonia.org.nz.

Donations and membership

The NZDPN is a Health Promotion Association registered with the New Zealand Charities Commission (Registration: CC10565). As well as encouraging research into dystonia and promoting awareness of our condition, our mission is to provide information and support to all those affected by dystonia. We are a 'grass-roots' organisation. Most of our leaders have dystonia themselves, and we are entirely reliant on donations, membership contributions and other charitable grants.

The Network invoices members once each year, in February, for the Annual Subscription.

Membership is $35 and applies to the calendar year in which the payment is made.

Receipts are issued for amounts of $100 and over, and otherwise on request. Any donations are tax-deductible because of the Network's status as a registered charity.

The Treasurer will issue donors with an official receipt which can be submitted to the IRD at the end of the Financial Year.

Internet Banking details are:

NZ Dystonia Network
Westpac, Takapuna
03 0275 0041784 000

Please complete the 'Code', 'Reference' and/or 'Particulars' fields with your name, and whether the deposit is for donation, seminar or membership payment.

Mission Statement: Our 3-fold mission is:

- To support dystonia patients with information, advice and networking opportunities
- To increase awareness about dystonia - both among the medical community and the public
- to encourage and facilitate research, with the aim of seeking better treatments, prevention, a cure

Disclaimer: Nothing in this newsletter is intended to serve as medical advice on dystonia. The NZDPN recommends that you consult your own doctor(s) and other health professional(s) regarding your diagnosis and treatment.

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New Zealand Dystonia Patient Network Inc.
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