National and international news and views
Networking Dystonia patients

New Zealand Dystonia Patient Network Inc.

News+Views

Newsletter 86 – November 2019

SAVE THE DATE: Saturday 16 May 2020

Brentwood HOTEL

Next year's Seminar will be held in Wellington at the Brentwood Hotel on Saturday 16th May. As usual the Seminar will be followed by the Annual General Meeting. More details will be available in the first newsletter of 2020, and on the NZDPN website - www.dystonia.org.nz.

Brentwood Hotel is located 2.6 km from the Wellington International Airport and 6 km from the city centre. If you wish to stay on-site over the weekend please book directly with them;

Location: 16 Kemp Street, Kilbirnie, Wellington
Phone: 0508 273 689 (0508 BRENTWOOD)
Email: reservations@brentwoodhotel.co.nz
Website: https://www.brentwoodhotel.co.nz

From the Chair – Alison Fitzpatrick

Greetings everyone! I hope you all managed to get through winter and are now enjoying the thought of spending time outdoors over the summer months ahead. I recently subscribed to a music streaming service, so everywhere I go I have musical accompaniment; both indoors and outdoors. This is a great novelty and it makes my daily walk very enjoyable. I find that certain types of music, especially full-on jazz like Dave Brubeck or Miles Davis, really seem to help my dystonia symptoms. I think it is the syncopated beat that makes me want to step out, but it also soothes the urge to move in a disordered way. This is possibly a placebo effect but if it helps the Dystonia, I'll take the improvement. Does anyone else find that music helps their Dystonia? - Let us know.

Letters to the Editor

We welcome your comments. Please forward any suggestions on how we can improve on what we currently publish to Jayne Lewington Lovell at jllovell@gmail.com.

Retirement Announcement

Our long-serving Network Manager, ***Philippa Hooper,*** retired from the position in August this year. Philippa was one of the founding members of the NZ Dystonia Network, and served as our Treasurer initially.

She was a tireless advocate for the Dystonia community, particularly in the area of family disability, where she was often contacted by parents of young children, seeking help and advice when their young ones were diagnosed with early-onset generalised dystonia. Philippa was driven by her determination to achieve the best outcomes for her younger son, Toby - seeking answers from the medical community to better cope with and understand his condition.

More recently Philippa served as our Network Manager. In this role she became the primary telephone contact person for the Network. She also liaised with Lotteries to help us accomplish our fund-raising goals. Philippa commissioned many of our publications, including our series of pamphlets that we send out to new members when they join and to healthcare providers on request. We will miss Philippa's involvement. Always cheerful, she had vast institutional knowledge and knew many of our members in person.

We all wish you well in your retirement Philippa!

Highlights from the 2019 Rotorua Seminar

It is hard to believe this is our last newsletter for the year! This edition will include presentations from Dr's Mark Simpson and Chris Lynch; plus DNA news of their Awareness Week in Sydney.

Seminar Presentation – Dr. Mark Simpson

Mark's talk focused on a medical perspective of dystonia. This included a historical background, the role of genetics and treatment options.

A brief history

- A psychogenic disorder? Says more about doctors than dystonia
- Hermann Oppenheim (1911) identified *dystonia musculorum deformans* and

early-onset generalised torsion dystonia

- Denny-Brown (1960) discovered that selective lesions in monkey brain produced dystonia
- Marsden (1976) proposed writer's cramp, torticollis and blepharospasm were all forms of dystonia
- Tsui (1985) reports the use of botulinum toxin in cervical dystonia
- DYT1 gene discovered in 1997

Genetic causes of dystonia

Genetics are a relatively new area of research. Mark informed us that twenty-three gene types have been identified so far, and that the most is known about DYT-1, DYT-5 and DYT-11.

Primary (DYT-1) dystonia was formerly called Oppenheim's dystonia or *dystonia musculorum deformans*. Symptoms generally begin in the first decade and present in an arm or leg, then later portending more rapid spread.

Dopa-responsive dystonia (DYT-5) is a rare, autosomal dominant disorder with onset in childhood. There is usually dystonic posturing of the foot which worsens as the day progresses. The disorder responds well to levodopa (hence the name), and thus, all patients presenting with early onset dystonia warrant a trial of levodopa.

Myoclonus-dystonia (DYT-11) is an autosomal dominant disorder, with an onset of symptoms before twenty years of age. Tends to involve arm and neck, and with shivering myoclonic jerks. DYT-11 is very responsive to alcohol.

Treatment Options

Botulinum toxin injection is the treatment of choice for blepharospasm, spasmodic dysphonia, torticollis, and focal dystonia of the hand. Side effects are minimal and the duration of benefit is typically 3-4 months.

Deep brain stimulation (DBS) is an important therapy, but careful patient selection is critical as this is not a treatment for all dystonia types; it does work well in DYT1 dystonia.

The ABCs of Pharmacotherapy for Dystonia

Anticholinergic – **B**aclofen - **C**lonazepam

Trihexyphenidyl, baclofen and clonazepam are most common in treating dystonia. In severe cases a combination of agents may include; tetrabenazine, levodopa, dopamine agonists and anti-epileptics.

In summary

- Dystonia is complicated
- Not every doctor knows about dystonia
- The genetics is complex
- Treatment is difficult
- Understanding and education is key

Seminar Presentation – Chris Lynch

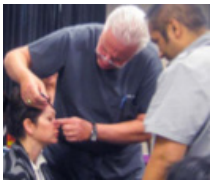
Dr. Chris Lynch shared experiences gathered over thirty years of working in medical Botox clinics. Chris is Clinical Director at the Midlands Botulinum Toxin Clinic, and, acknowledges that his hands-on approach is based upon things learned from; neurology colleagues, teachers, the industry (Allergan), and most importantly his patients.

Some of the learning effects mentioned were;

1. Feedback - how did it go and how can I make it better?
2. Nurse – assists with running of clinic and is a point of contact for patients
3. Pain – applying an anaesthetic cream before injecting
4. Aesthetics – adjusting treatment to remedy things like droopy eye

Chris is also involved in teaching initiatives and professional development. An example of this is an annual Botox® workshop for neurologists.

The workshop is a chance for those in the medical dystonia community to meet once a year so they are not working in isolation. This a team approach to improving service delivery for patients. They are also useful to patients as they get a longer appointment time and are part of the discussion of diagnosis treatment; trainees gain valuable experience from both senior injectors and patients; and lastly everyone contributes to the teaching and practice methods for future injectors.



Update from Australia – DNA

As reported in the last newsletter, the DNA had their Dystonia Awareness Week in September. One highlight was a half hour program on ABC Radio called 'the Health Report'. This is an interesting interview with emphasis on the personal stories of two dystonia patients: Robyn and Paul. There is also medical expertise from neurologists Victor Fung & Stephen Tisch; and researcher Lynley Bradnam. The link for the podcast is;
https://www.abc.net.au/radionational/programs/healthreport/living-with-the-mental-weight-of-dystonia/11491220

'Dystonic Storm" – A Poem

On a clear day I'll flippantly
tell you that 'Dystonic Storm'
would make a great name for a
rock band - and it would

But when the mercury falls and
winds blow anywhichway my body
barometer rides the Beaufort Scale
from 1 to 12 – and back again

The band of pressure over head
flashes lightning very frightening
thunder rumbling causes mumbling
stumbling – and confidence crumbling

Am losing words in my distress
Mayday Mayday SOS; Calling all
cars Call out the coastguard Call
out anybody - and my mother

A signal comes on the breeze
tap, tap, the mercury is rising
like my hope that I can face a
new day - and carry on

On that new day I'll flippantly
tell you that 'Dystonic Storm'
would make a great name for a
rock band - and it would

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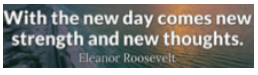

Movement Disorder Blog: Rehabilitation as a Therapeutic Approach for Dystonia

Authors: Amit Batla, *MBBS, MD DM,* Lynley Bradnam, *PhD,* and Teresa Kimberley, *PhD.*
In this blog the authors discuss the status of employing a rehabilitation approach for therapy of dystonia.

• What limitations exist in current treatment?

Dr. Batla:
Current treatments for dystonia are mainly symptomatic and are not curative. First are the oral treatments such as anticholinergic, dopaminergic, and GABAergic medications; whose efficacy is compromised by side effects. For focal dystonia, botulinum neurotoxin injections are the main treatment. These are painful and require clinic visits, usually every three months. There are also some forms of dystonia that are more responsive or resistant to the regime. Deep brain stimulation (DBS) is an important therapy but appropriate utilisation is limited by high costs, patient eligibility and regulatory challenges (the FDA has approved DBS for dystonia only under a humanitarian disease exemption category).
Dystonia is also associated with non-motor symptoms; however, these features are often under-recognised and undertreated. Current treatments are not useful to reduce symptoms like pain, anxiety, and depression which greatly affect the quality of life of dystonia patients.

• Is there a role for rehabilitation in dystonia?

Dr. Bradnam:
Definitely! Our recent research has proven people with dystonia have additional functional deficits, some affecting balance control. Some patients may have a fear of falling and visual difficulties that significantly reduce the quality of life. Physical rehabilitation should be utilised from a holistic perspective, not just targeted at the dystonic body part. For example, most therapeutic interventions for cervical dystonia to date have focused on exercises aimed at reducing the neck impairment, but have had limited success. It is necessary to encourage people with dystonia to engage in physical activity to maintain/improve their physical and psychological health and well-being.

Dr. Kimberley:
There is significant disability associated with focal dystonia due to pain and impairment, reduction in participation of daily activities, employment, loss of self-confidence, and fatigue. These can be addressed with physical therapy. Many small-scale studies have explored the potential benefits of strategies such as

transcutaneous electrical nerve stimulation (TENS), Braille training, movement practice and vibration-based training. There are varying degrees of success with the application of these strategies. Improvements have been observed in terms of reduced disease severity, quality-of-life, and motor performance. Physical therapies have a promising role in retraining more normal movement patterns and postures within the context of daily activities such as walking, running, reaching and grasping, and moving from one place to another.

• **Which techniques will be most effective?**

Dr. Bradnam:

We have to think about the brain network sub-serving dystonia and design interventions to target that network. Exercises serve to simulate signals from muscles to the brain, which, if delivered incorrectly, can overload the dystonic brain already processing sensory information in a dysfunctional way. It is important not to utilize a 'one size fits all' approach. Assessment of the main motor (tremor) and non-motor (fatigue), symptoms may give clues as to the parts of the neural circuitry that is dysfunctional in that individual; and this can then be used to guide interventions like exercise.

Dr. Kimberley:

It is an essential that movement disorder clinics recruit physical and occupational therapists, as well as investing in their training for effective treatment strategies for people with dystonia. This has the potential to extend the benefit of botulinum toxin and provide an alternative for those that seek it. We also need to recognize that movement practice (through rehabilitation) is a powerful driver of neuroplastic change in the brain. In closing, rehabilitation has a promising role as an adjunct treatment; but its potential has not been fully explored. A multidisciplinary program that addresses the many facets of dystonia in the patient as a whole, could fill the existing gaps, and realize the much needed therapeutic success in this complex disorder.

For the full article go to the MDS website;

https://www.movementdisorders.org/MDS/Scientific-Issues-Committee-Blog/Rehabilitation-as-a-Therapeutic-Approach-for-Dystonia.htm

Governance of the NZDPN

The Executive Committee is elected each year at the Annual General Meeting. The Committee for 2019/2020 is;

Chair: Alison Fitzpatrick

Secretary: Desiree Sargon

Treasurer: David Barton

Committee Members: Jayne Lewington Lovell, Roger Terry, Dave Mitchell

NZDPN email: 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. All 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 $30 per annum 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

ANZ BANK, WAIKANAE. 06-0577-0110415-00

Please complete the 'Code', 'Reference' and/or 'Particulars' fields to let us know your name, and whether a deposit is a donation or membership.

If you prefer to send a cheque our address is:

The Treasurer, NZDPN, PO Box 34 259, Birkenhead, Auckland 0746

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.

www.dystonia.org.nz

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