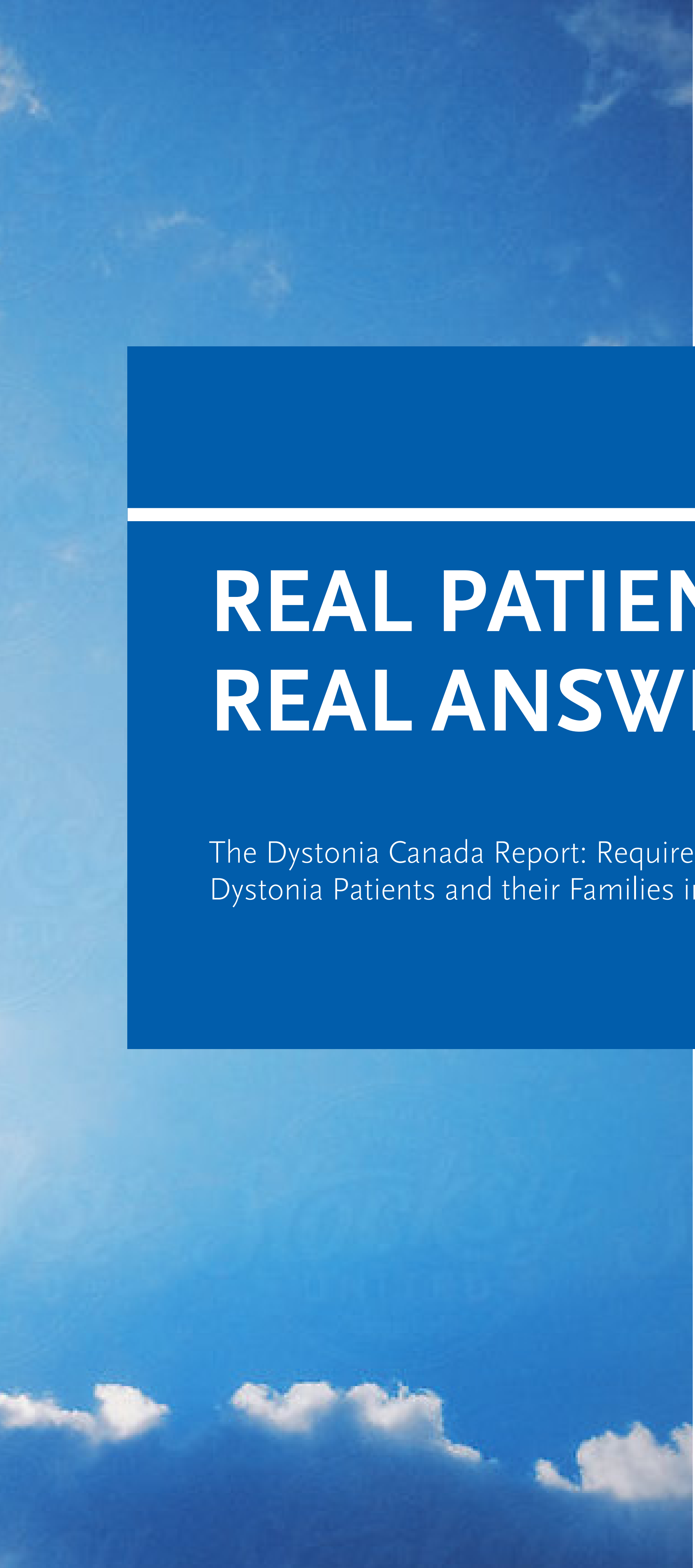

REAL PATIENTS.
REAL ANSWERS.

The Dystonia Canada Report: Requirements for Dystonia Patients and their Families in Canada.

Dystonia
Medical
Research
Foundation
Canada

Fondation de
Recherche
Médicale sur la
Dystonie
Canada

serving all dystonia-affected persons
désservant toutes personnes atteintes de dystonie

> People really suffer from this disease. Thank you for helping to raise awareness for a condition few people even know about.
>
> – *Survey Respondent*

Dystonia Medical Research Foundation Canada

The Dystonia Medical Research Foundation (DMRF) Canada is the only organization that is dedicated to supporting the 50,000 patients in Canada that suffer from dystonia, a neurological movement disorder that affects the nervous system. Abnormal signaling from the brain causes muscles to tighten and twist involuntarily. There are several forms of dystonia and dystonia may be a symptom of many diseases and conditions.

For over 40 years, DMRF Canada has fought for those impacted by dystonia, working toward better treatments and a cure, funding research, providing advocacy and support, and raising awareness of this devastating disorder.

Our role as a charitable organization is to give those in Canada suffering from dystonia with a clear and strong voice. What better way to do this then to allow our patients to speak out for themselves? This document represents the honest responses about the experiences, feelings and thoughts experienced by real patients in Canada.

The results are troubling. But they also provide hope. There is much to be done to improve the day to day lives of patients living with dystonia in Canada. Please join us and be a part of our effort to find better treatment, and a cure. Help us with our aim to educate the public, key stakeholders, and the dystonia community on behalf of dystonia patients in Canada.

Notice of Copyright and Intellectual Property

Our thanks to André-Barbeau Movement disorders unit, CHUM; Our thanks to Dr. Sylvain Chouinard, MD, FRCPC, Co-director, André Barbeau Movement Disorders Unit, CHUM, for his review and input into the development of this publication.

Data from the Survey: This data was collected between August 1, 2016 and December 31, 2016. Specific research data from the survey may be available for further investigation. If you are interested in reviewing this survey data please submit a formal request to Stefanie Ince, Executive Director at the Dystonia Medical Research Foundation Canada. Please contact ***info@dystoniacanada.org*** for more information.

The DMRF Canada is a registered charity whose mission is to advance research for more treatments and ultimately a cure; to promote awareness and education; and to support the needs and well being of affected individuals and families. ***www.dystoniacanada.org***. Phone: 416-488-6974.

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Summary of the Dystonia Canada Survey:

The focus of the Dystonia Survey was to provide a comprehensive evaluation of the experiences, fears, feelings and thoughts of Canadians living with dystonia, to help the DMRF Canada develop a long-term strategy for better supporting this patient population.

This pioneering research study was the first of its kind in Canada. Close to 900 surveys were completed by dystonia patients. After removing responses from individuals from outside of Canada, as well as duplicates, we had over 600 responses from dystonia patients across Canada, collected over a four-month period. Responses were provided through an online survey that was promoted electronically and via social media, in the DMRF Canada Fall 2016 Newsletter, and via printed brochures that were distributed at movement disorder clinics across the county in both official languages. A note that the results of the survey may be influenced by the fact that one of the main channels used to recruit survey participants was to distribute surveys to movement disorder clinics, where patients are seeking treatment by neurotoxin.

Survey respondents came from Canadians across ten (10) provinces and the Yukon Territory; it represented all ages, genders, and over twenty forms of dystonia.

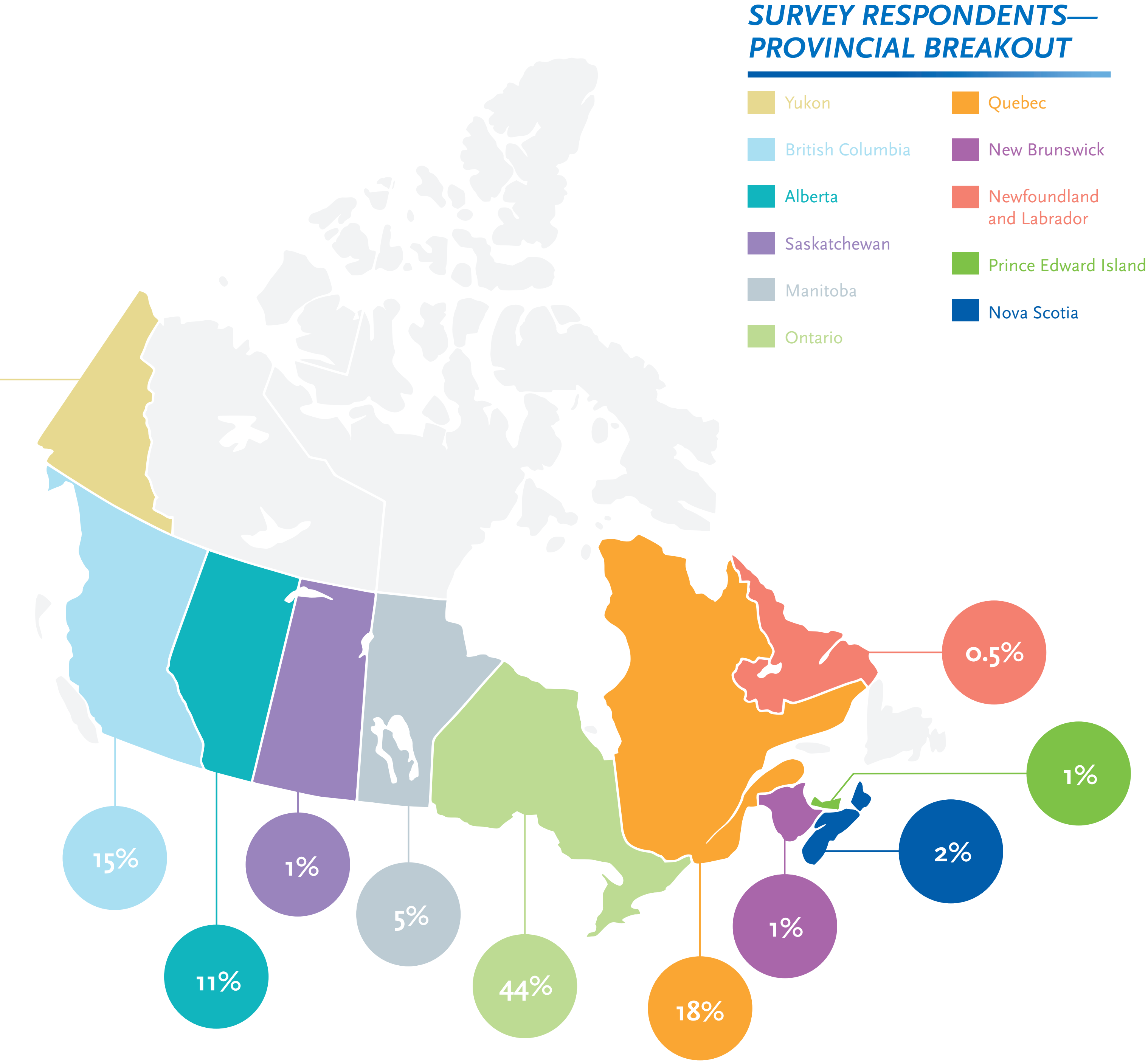
0.5%

Real Patients: Survey Respondents

A total of 613 surveys were included in English and French . 79% of survey respondents were female and 21% were male. 41% of survey respondents were between the age of 41 and 65, and 31% were over the age of 65. Only 9% were under the age of 41.

> “Cervical Dystonia has changed my WHOLE life. Everything is a challenge; from personal grooming to social events, to going grocery shopping.”
>
> *– Survey Respondent*

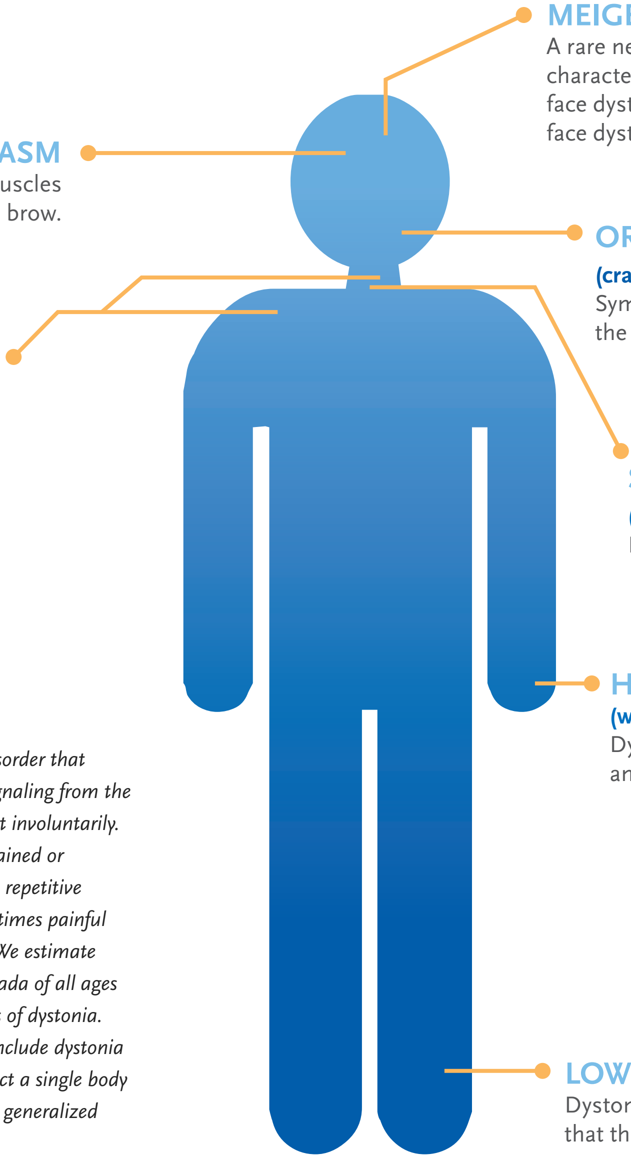
Dystonia Voices Across Canada


SURVEY RESPONDENTS—PROVINCIAL BREAKOUT

Forms of Dystonia

FOCAL DYSTONIA:

The most common form of dystonia, occurring in adulthood. Focal dystonias affect a specific area of the body. Most focal dystonias are primary (meaning that it is the only neurological symptom and presumed to have a genetic component), though secondary cases are documented. Focal dystonia may affect muscles of the eyes, mouth, vocal cords, neck, hands, and feet. The following are forms of focal dystonia according to the body part that is affected:



MEIGE SYNDROME

A rare neurological movement disorder characterized by a combination of upper face dystonia (blepharospasm) and lower face dystonia.

BLEPHAROSPASM

Dystonia that affects the muscles of the eyelids and brow.

OROMANDIBULAR DYSTONIA

(cranial dystonia)

Symptoms include forceful contractions of the face, jaw, and/or tongue.

CERVICAL DYSTONIA

(spasmodic torticollis)

Dystonia that affects the neck and sometimes the shoulders.

SPASMODIC DYSPHONIA

(laryngeal dystonia)

Dystonia that affects the vocal cords.

HAND DYSTONIA

(writer's cramp)

Dystonia of the fingers, hand, and/or forearm.

LOWER LIMB DYSTONIA

Dystonia of the leg, foot, and/or toes. A note that this is extremely rare in adulthood.

What is Dystonia?

Dystonia is a neurological movement disorder that affects the nervous system. Abnormal signaling from the brain causes muscles to tighten and twist involuntarily. These muscle contractions could be sustained or intermittent, and cause abnormal, often repetitive movements, postures or both. It is sometimes painful and may interfere with daily activities. We estimate that dystonia affects over 50,000 in Canada of all ages and backgrounds. There are many forms of dystonia. Dozens of diseases and conditions also include dystonia as a major symptom. Dystonia may affect a single body area (e.g.: face, neck, hands, feet), or be generalized throughout multiple muscle groups.

MUSICIAN'S DYSTONIAS:

Task-specific dystonias sometimes given this name because constant use of various muscles and movements can contribute to dystonia among musicians related to their performance.

EARLY-ONSET GENERALIZED DYSTONIA (DYT1 and non-DYT1):

Characterized by twisting of the limbs and torso.

DOPA-RESPONSIVE DYSTONIA:

Refers to a group of dominantly inherited dystonias that respond to a medication called levodopa.

MYOCLONUS DYSTONIA:

A hereditary form of dystonia that includes prominentmyoclonus symptoms.

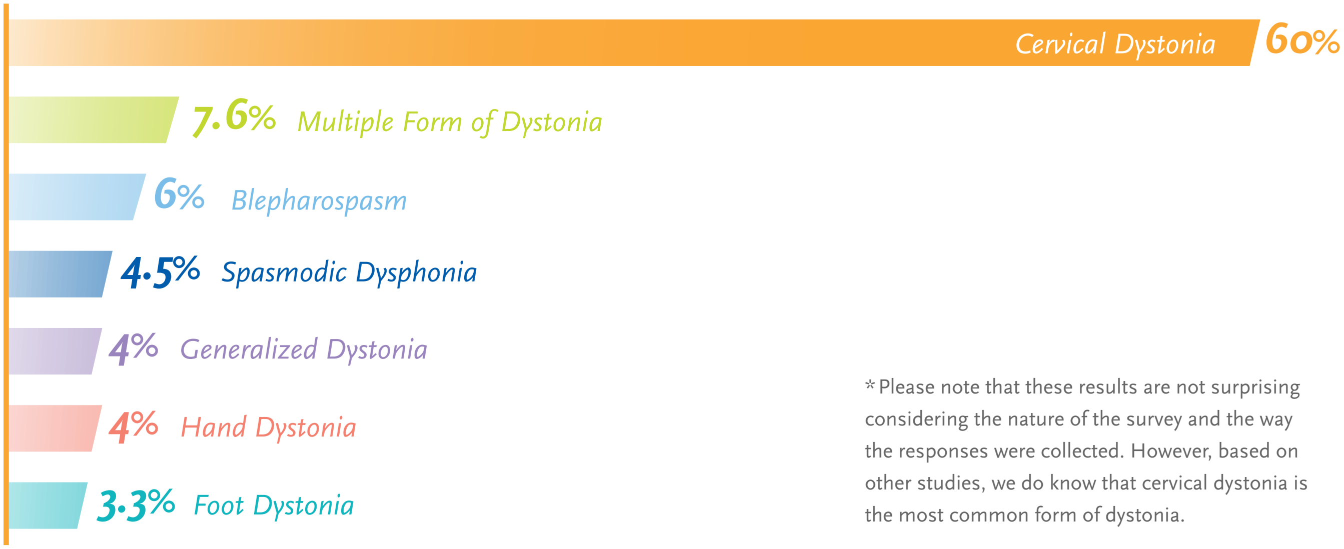
PAROXYSMAL DYSTONIAS AND DYSKINESIAS:

Involves episodic movement disorders in which abnormal movements occur only during attacks.

For more information on the various forms of dystonia visit: ***www.dystoniacanada.org***.

Patients Respond: Forms of Dystonia

The majority of survey respondents confirmed that they had cervical dystonia (*60% of respondents*),* but other forms of dystonia were listed, including:



* Please note that these results are not surprising considering the nature of the survey and the way the responses were collected. However, based on other studies, we do know that cervical dystonia is the most common form of dystonia.

Dystonia Treatment — The Use of Neurotoxin

What Is Botulinum Neurotoxin Therapy?

Botulinum neurotoxin (neurotoxin) is a medical treatment derived from bacteria called Clostridium botulinum. Several neurotoxins are used as injected medications to treat dystonia and other disorders. This class of medication has been approved for use in Canada for nearly 30 years.

How does Neurotoxin work as a Dystonia Treatment?

When neurotoxin is injected into muscles affected by dystonia, it blocks the neurotransmitter chemicals that signal the muscles to contract and spasm excessively. The muscle is weakened and therefore relaxes, reducing the dystonia. Prior to the introduction of neurotoxin during the 1980s, there was essentially no treatment for focal dystonias, and generalized dystonias were managed with oral medications and brain surgery.

Who Is Using Neurotoxin?

When we looked at those individuals who were using a neurotoxin based on the form of dystonia they had, we noticed some clear differences. 89% of individuals who suffer from blepharospasm currently receive botulinum neurotoxin, while only 32% of those patients who suffer from generalized dystonia currently receive this form of treatment.

It is interesting to note that 32% of the people that suffer from generalized dystonia are receiving this form of treatment as well. Generalized dystonia was more often treated in the past by oral medications only, and the survey pointed out that neurotoxin injections could be used as an add-on treatment for generalized dystonia.

We were pleased to see that access to neurotoxin treatment for dystonia was available across Canada. However, there were clear differences in terms of the percentage of people using this form of treatment by province. This could be due to a bias in the study or differences in accessibility across the country.

62%

of survey respondents confirmed that they currently receive botulinum neurotoxin injections for their dystonia.

It is important that dystonia patients consider neurotoxin as a possible treatment method in all provinces. There appears to be inconsistency regarding the percentage of people using this method of treatment in communities across the country.

> In 2017, Botulinum Neurotoxin Therapy is still the best treatment for focal dystonia but the survey has clarified that this is not always a perfect form of treatment. We definitively need to continue to work to find a cure
>
> *– Dr. Sylvain Chouinard*

Dystonia Treatment —The Use of Neurotoxin

Frequency of botulinum neurotoxin injections among patients.

63%
receive their injections every 12 weeks

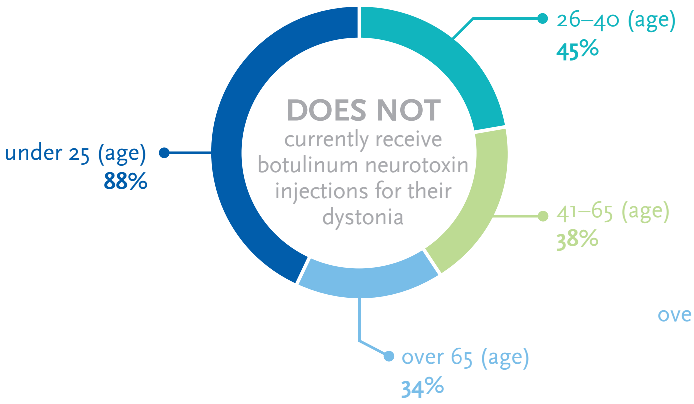
19%
receive their injections less frequently than every 12 weeks

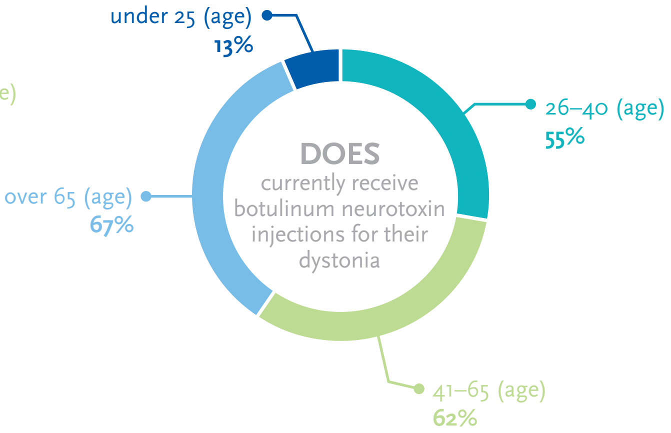
18%
are being injected more frequently
either every 6 weeks, every 8 weeks, or every 10 weeks

> “These results represent a recent change in the practice. The concept of flexible interval injections is starting to emerge. We would not have seen these results in a survey ten years ago. At that time, everybody was treated every 12 weeks—or even less frequently.”
>
> *– Dr. Sylvain Chouinard*

Neurotoxin Use by Age

A higher percentage of survey respondents over the age of 41 were users of neurotoxin than those under the age of 41. For those individuals under the age of 25, only 12.5% were users of neurotoxin. For individuals between the age of 26 and 40, 55% were users of neurotoxin, which is lower than the overall average of neurotoxin use. This finding only clarifies that younger patients have more generalized dystonia and are therefore less likely to receive neurotoxin injections.





How Dystonia Treatment is Covered

For the most part dystonia patients who receive neurotoxin treatment are covered in part or in full, either by private insurance or by a provincial plan. 33% of survey responses confirmed that they have private insurance, but that it doesn't pay for 100% of coverage. 29% confirmed that they are 100% covered by their provincial plan, and 24% confirmed that they have private insurance that pays 100% of their medication cost.

33%
have private insurance, but it doesn't pay for 100% of coverage.

29%
are 100% covered by their provincial plan

24%
have private insurance that pays 100% of their medication cost

The question we did not ask, but will explore further in the future, is how a person's medical coverage may have impacted their choice to seek out appropriate treatment methods. From the perspective of the DMRF Canada, it is important that all dystonia patients have access to the same coverage for the best treatment for their dystonia.

The Real Truth — Dystonia Patients Are Not Getting the Help They Need

Satisfaction with Neurotoxin

For all individuals who use neurotoxin, only 13% describe their dystonia as 'under good control' on the scheduled day of their injection appointment. 38% describe their dystonia as "manageable" on the day of their injection appointment. 24% describe their dystonia as 'troublesome' and 25% describe their dystonia status as inconsistent, saying: "*my dystonia status seems to be different each time I go for treatment, it may be okay or it may be troublesome*".

13%
under good control

38%
manageable

24%
troublesome

25%
inconsistent

Most Common Concerns about neurotoxin

For those individuals who are currently taking a neurotoxin, we asked *"What concerns you the most about your neurotoxin treatment?"* The most common answers were:

26% The cost of the medication

26% The safety of the medication

19% The pain of the needle

We also included an "Other" category and found that several responses could be grouped into general categories that were not listed on the survey:

8% Concern with long-term effects and losing effectiveness over-time

4% Concern that it is not working as much as it should or at all

3% Concern it will make symptoms worsen or cause other complications

DMRF Canada encourages patients and their health care providers to have in depth conversations about how to improve their overall satisfaction with their treatment and results.

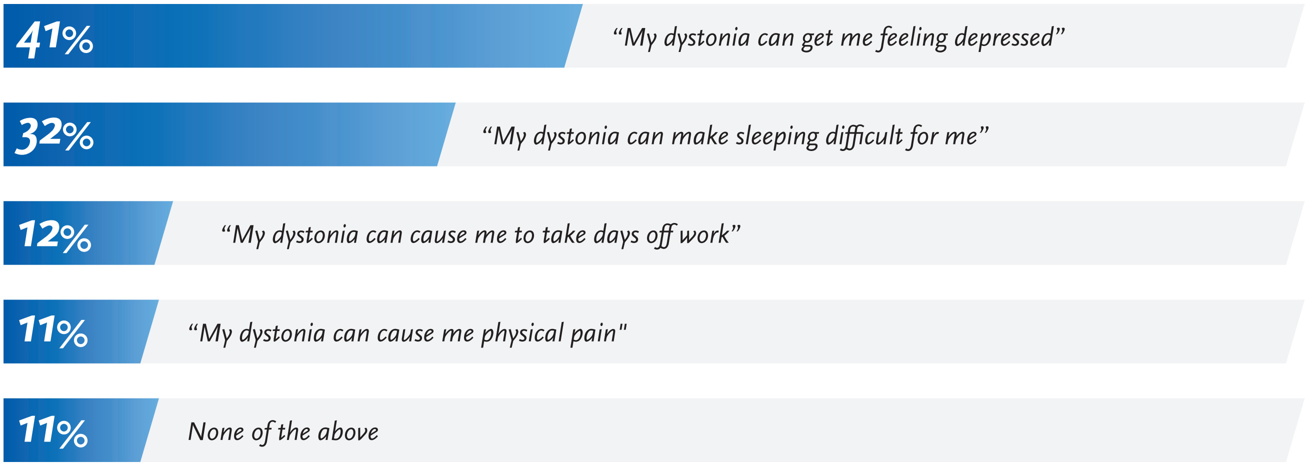
Patients Are Looking For More Answers

Despite having some treatment options, including neurotoxins, other oral medications, and in some cases, surgery, many patients are looking for news on alternative treatments for their dystonia. When asked whether they were interested in learning more about changes in research findings or available treatments, when those treatments became available, 88% confirmed they would. DMRF Canada has taken note of this finding. We will continue to work very hard to identify new research findings, new treatment methods, and new information about existing treatments.

The Real Truth — A Painful Existence in Many Forms

We asked survey respondents who were taking a neurotoxin, *"As the effect of your injections begins to wear off, which of the symptoms start to re-emerge and affect your day-to-day life and activities"* Below is a breakdown of their responses. Please note that respondents could provide multiple answers.

We asked How Do Your Symptoms Affect Your Day-to-Day Life and Activities?



Depression

The most common symptom that starts to emerge for those neurotoxin users when their injections began to wear off was depression. This was reinforced in a follow up question that we asked later in the survey; *"What is the biggest challenge you face with your dystonia?"*

The most frequent answer to this question was some form of anxiety and/or depression — this represented 32% of all responders.

32%

of responders feel the biggest challenge they face with dystonia is anxiety and/or depression.

Some patients broke this down further, and specified that this was due to social anxiety and embarrassment to be around people when visibly disabled (13%). Others noted that they felt depressed about losing their ability to do basic tasks they once took for granted (14%).

We analysed the relationship between anxiety and depression against various forms of dystonia. People who listed anxiety and depression in general as their greatest challenge were evenly distributed across different dystonia types. However, depression due to loss of abilities was most common among individuals with generalized dystonia (20%), cervical dystonia (16%), and hand dystonia (13%).

Social anxiety was most significant among patients with dystonia affecting the face, head, or voice. For patients with facial/hemi-facial dystonia (21%), patients with meige syndrome (20%), patients with cervical dystonia (15%), patients with oromandibular dystonia (14%) and patients with laryngeal dystonia (26%) highlighted social anxiety as their greatest challenge.

"I feel I have no where left to turn"

"A feeling of isolation"

"Every aspect of day-to-day living and raising a 5 year old with this monster named Dystonia"

The Real Truth — Everything Can Be a Challenge

Posture, Mobility Coordination, Motor Control

In some instances, the challenges were specifically related to the type of dystonia an individual had. For example, of the people who complained of vision problems, 77% of them were diagnosed with blepharospasm. The most common challenge for patients with focal foot and focal hand dystonia was issues related to posture, mobility, and coordination. (58% of focal foot patients, and 42% of focal hand patients).

"I am often self-conscious about the irregular postures and movements caused by the disorder, which can in turn magnify the symptoms."

Pain

When asked this question, many people confirmed that pain is the biggest challenge of living with dystonia.

"The horrid pain that comes with it. Dystonia is stopping me from living"

Financial Stress

Employability and Financial stress was named as one of the other challenges an individual faced due to their dystonia. Of those individuals who take a neurotoxin and who were under the age of 65, 14% said that their dystonia could cause them to take time off work. People between the ages of 41 and 65 reported financial issues as their greatest concern (78%) more often than other age groups.

Not Enough Support

Overall, there is a feeling that there is not enough support for dystonia patients in Canada – both within the medical community, as well as within the general public. There is a feeling of frustration and a call on the DMRF Canada to help raise awareness for dystonia to ensure all dystonia patients are getting the medical help they need.

> *"I think there is very little public info on how debilitating this condition can be. The diagnosis is very empiric and almost nobody knows about dystonia"*

Moving Forward with Hope

Clearly there are several opportunities available to better support the dystonia community in Canada, but the Dystonia Survey was an important first step that has provided us with a better understanding of the experiences, fears, feelings and thoughts of Canadians living with dystonia.

It also gave a chance to see that the dystonia community is incredibly brave, strong, and not backing down from their condition.

> ***"With research and time we will beat this crippling disease"***

> ***"I will overcome this disorder somehow..."***

> ***"Thank-you for showing interest in a condition that I often say I would not wish on my worst enemy."***

Living Well with Dystonia:

There is no single strategy for living well with dystonia that works for everyone. The following suggestions have been compiled from healthcare professionals, individuals with dystonia and support leaders:

Seek Out Expert Healthcare Providers:
It often takes a team of experienced professionals to diagnose and treat dystonia. This may include a movement disorder neurologist, physical therapist, psychiatrist, psychotherapist/counsellor and others.

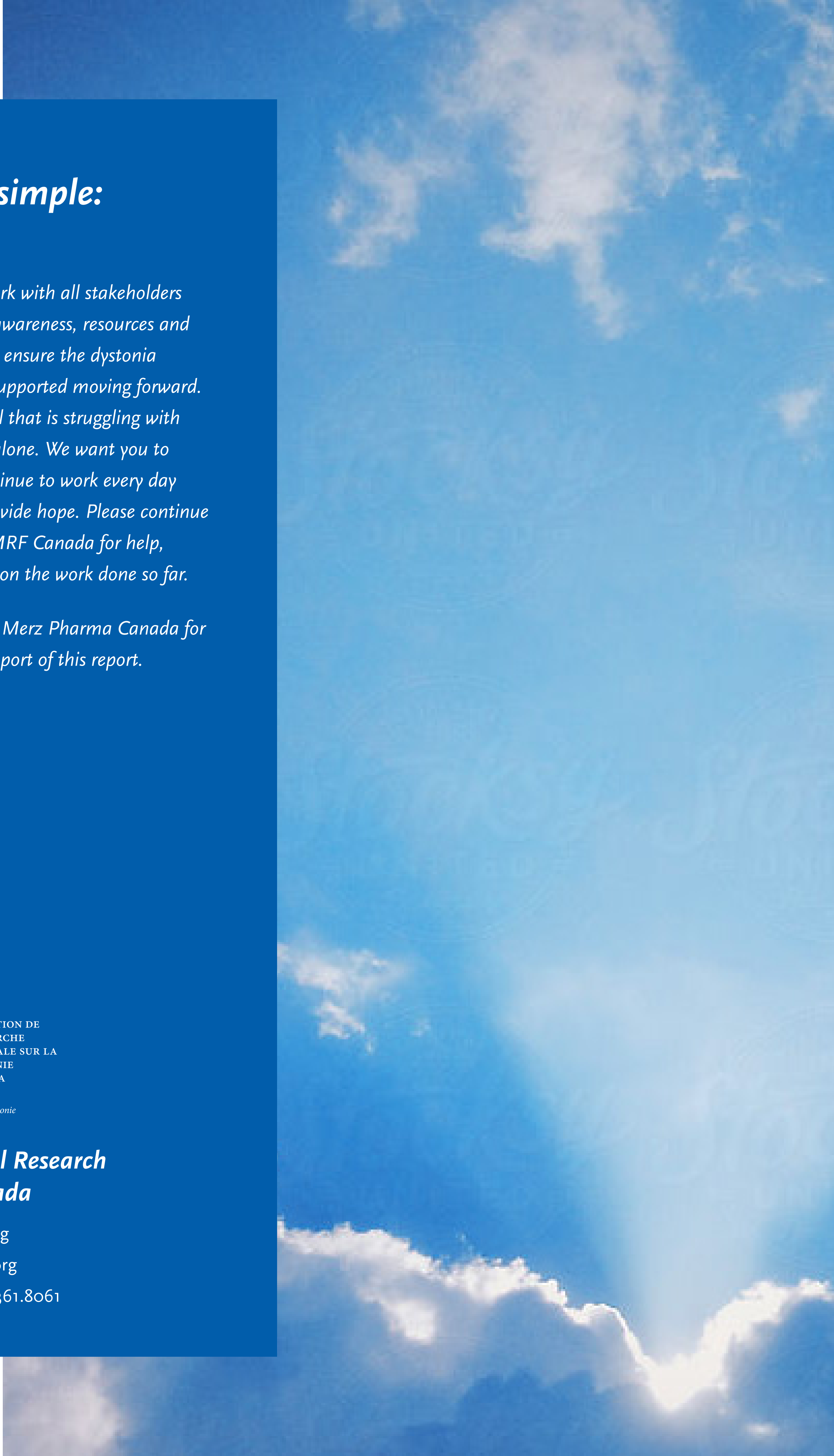
Actively Participate in Your Treatment:
Learn about dystonia. Make informed choices about your care with the input of your medical team.

Be Mindful of Stress:
Regular stress reduction and relaxation practices can have a positive impact on symptoms and overall well-being.

Remain as Active and Social as Possible:
It may be physically and/or mentally demanding to go certain places or complete specific tasks. Plan ahead, pace yourself and rest when needed.

Take Care of Your Relationships:
Resist the temptation to isolate from other people. If family or love relationships are strained, consider enlisting the help of a therapist to bring you closer to those you care about.

For more tips on how to live well with Dystonia vist:
dystoniacanada.org/patient-care-package.



Our goal is simple: To do more.

We will continue to work with all stakeholders to develop additional awareness, resources and education materials to ensure the dystonia patient population is supported moving forward. If you are an individual that is struggling with dystonia, you are not alone. We want you to know that we will continue to work every day to do more, and to provide hope. Please continue to reach out to the DMRF Canada for help, or to provide feedback on the work done so far.

A sincere thank you to Merz Pharma Canada for thier very generous support of this report.

DYSTONIA MEDICAL RESEARCH FOUNDATION CANADA

FONDATION DE RECHERCHE MÉDICALE SUR LA DYSTONIE CANADA

serving all dystonia-affected persons
désservant toutes personnes atteintes de dystonie

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