



Cerebral
Palsy
Research
Network

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The Dystonia in CP Toolkit

FROM DIAGNOSIS TO UNDERSTANDING

FIRST EDITION

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The issue of Dystonia in Cerebral Palsy (DCP) is critical to understanding what challenges face parents, caregivers, children and adults affected by DCP. As a parent of a young adult with DCP, and one of the co-founders of the Cerebral Palsy Research Network, I know it can be difficult to access information on this topic. Approximately 70 percent of people who have CP also have dystonia.¹ Currently, there are more questions about DCP than answers. Even though the current state of science about dystonia in CP is limited, a path forward is emerging regarding how to address it. Science takes time. At the CP Research Network we encourage scientists to use the most efficient and powerful methods to find the answers we desperately desire. As you read about dystonia in CP, take in the information at your own pace and as you feel comfortable. You may wish to discuss this important topic with your healthcare providers.



Please join us on our socials, in our community discussion space, and on our webinars for more information about cerebral palsy

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We welcome you and hope you learn some important information about dystonia in CP.

Sincerely,

A handwritten signature in black ink that reads "Michele Shusterman". The signature is fluid and cursive, with a large, expressive "M" at the beginning.

Michele Shusterman
Co-Founder of the CP Research Network,
Director of Education and Engagement

Foreword



"It was only when he received the diagnosis of CP with dystonia (DCP) that he was able to get the help and treatment he needed."

-Sean's mom Jill

The Dystonia in CP Toolkit was developed after CP Research Network leadership asked community members about areas of research they felt were unsupported or there were treatments lacking. We have created this resource and other related educational materials to help you better understand DCP and the state of research and treatments. One parent caregiver expressed the following concern,

"Dystonia seems to affect a lot of people in our population, but I never hear anything about it."

This parent's statement led to conversations among community members and researchers about dystonia in cerebral palsy. There was widespread agreement that this was a topic of great concern for the CP community and the clinicians who treat them. The consensus was that dystonia in CP had been largely overlooked as a condition that affects many people with CP and causes symptoms that have not been adequately addressed.

The following is a portion of a community member's experience with DCP. Many people with CP face similar challenges as Sean and his family from receiving a diagnosis to identifying the most effective interventions and treatments for their symptoms.

SEAN'S STORY

Sean was diagnosed with dystonia at age 18 after years of worsening symptoms. He had seen specialists for his CP in the past and the family had a good understanding of the impacts and treatments for him during the early part of his life. Yet, in his teens, Sean's symptoms changed, and some part of his movement disorder began to impact all activities of daily living including eating, sleeping, sitting comfortably, personal care and emotional wellbeing. He could no longer drive his wheelchair nor use his computer. No one was able to tell them why or give them help until they connected with a movement disorders specialist who diagnosed Sean with dystonia.

What Is Dystonia?

“I experience it more with deliberate movements that take concentration.”

-Anonymous adult with CP

“I notice it when I am stressed—in a time sensitive way or with excitement—a lot more than when I am not stressed.”

-Anonymous adult with CP

Dystonia is a movement disorder in people with CP that causes slow, uncontrolled, involuntary movements that emerge by certain triggers.²

Types of dystonia in CP:

- Generalized dystonia: Dystonia is experienced throughout the body.
- Focal dystonia: Dystonia is limited to a certain part of the body.

Common triggers for dystonia include:

- Attempting voluntary movement.
- Performing difficult tasks.
- Feeling excited or feeling unwell.
- Having someone move your body.

Why do we need to know about dystonia in CP?

Parents, caregivers and people with CP need to know about dystonia because it is common among people with CP (approximately 70 percent have it), and there is often a delay in diagnosis.¹ Dystonia is also a common cause of pain in CP and when diagnosis is delayed then treatment is delayed.³

Dystonia can emerge within the first several months of life, but the average age of referral for dystonia-specific treatment is about 8 years.⁴ This gap means that many children with dystonia don’t receive a diagnosis or treatment for several years.⁵

Knowing you have dystonia is also important for physicians and therapists to effectively manage the symptoms that may be interfering with your daily activities and comfort. This situation is where you or your parent/caregiver can help. By bringing up the possibility of dystonia with your medical team, you may shorten the time it takes for you or your loved one with CP to access the care and treatment that can help.

How May Dystonia Impact a Person's Life?⁶

- 1 Dystonia can affect how the person moves and coordinates movement (including speech).
- 2 Dystonia can interfere with positioning for sitting and lying.
- 3 Dystonic postures and movement can be painful and interfere with daily activities.³
- 4 Dystonia can interfere with sleep.
- 5 Dystonia can result in the person using high amounts of energy and may lead to problems with nutrition because the extra muscle movements cause the body to use more energy.
- 6 Dystonic postures can create challenges with caregiving.
- 7 Dystonia can impact wellbeing.
- 8 Dystonia can be associated with orthopedic deformities.

What is Cerebral Palsy?

Cerebral Palsy (CP) is the most common form of movement dysfunction in childhood. It affects 2-3/1,000 children and affects a large adult population.⁷

CP refers to several types of movement disorders caused by an early brain injury or disturbance in neurological development. Although CP may affect different systems throughout the body, it is the difficulty and symptoms associated with creating and coordinating movement that distinguish cerebral palsy from other early injuries to the brain.

The symptoms of CP impact people differently, including the parts of the body that are affected and to what degree. Dystonia is the predominant form of tone in 15 percent of people with CP, but even with other primary types of CP, such as spastic forms, there is increasing awareness that dystonia is often present as well.⁸

Ask the members of your medical team about their observations of you or your loved one's movements, how they may impact them and what you can do to possibly improve any symptoms that may be bothersome.

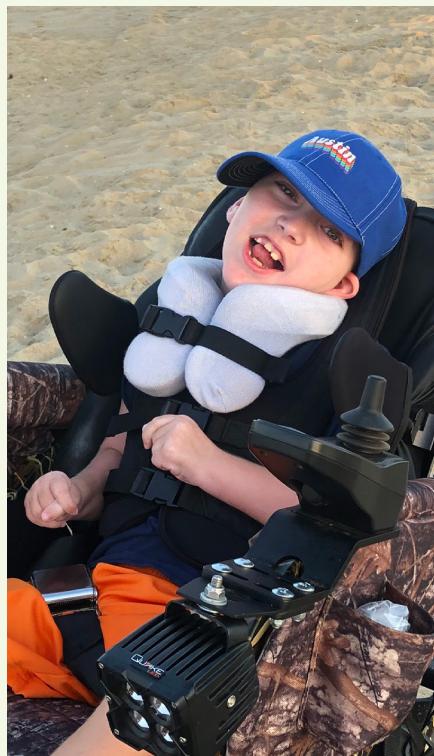


How Dystonia is Diagnosed

Dystonia in CP is often diagnosed through an extensive history in combination with a physical exam. **It is not typically diagnosed during one examination.** To assess dystonia, physicians may use tools such as the Barry-Albright Dystonia Scale, and the Hypertonia Assessment Tool.⁶ Clinicians can find resources about dystonia under the Care Pathways section of the [American Academy for Cerebral Palsy and Developmental Medicine \(AACPDM\) website](#). The CP Research Network is now strategically working toward having a dystonia assessment routinely included in the CP neurological examination.

The diagnosis of dystonia is much different than the diagnosis of spasticity. If the individual has more than one movement condition present such as spasticity and dystonia, it may be hard to identify the dystonia. This is why research and education are so vital. Since different movement disorders respond differently to treatments, receiving an accurate diagnosis is important.

Your healthcare team will complete an assessment to understand the severity of dystonia and how it is impacting you/your child's life. This assessment may include looking at different movements, stretching muscles to measure stiffness, and asking questions about how dystonia impacts daily activities, pain/comfort, and sleep.





Challenges to Diagnosing Dystonia

- 1 Clinicians (doctors, physical therapists) do not always look for it because they may not be educated in how to screen for it.⁹
- 2 Sometimes dystonia may be present on an exam and other times a clinician may not see it or it may not even be present.¹⁰
- 3 Dystonia often requires multiple evaluations and can't always be diagnosed with just a picture or a static moment in time.¹¹
- 4 Because people with cerebral palsy often don't know that they have dystonia, they don't bring it to the attention of their clinicians in a direct way. For example, there are certain conditions such as reflux, and sleeplessness that can worsen dystonia and dystonia may also increase these problems.^{6,11}
- 5 Dystonia in cerebral palsy can be difficult to identify especially when there are other movement disorders present. For example, a person with cerebral palsy may have spasticity that changes how the dystonia impacts their movements and how the dystonia appears to the evaluating medical team. This complication is very different than individuals who have dystonia but who don't have CP. In those cases, the changes associated with dystonia are obvious.¹²

Treatments

Most of the current recommendations for treatment are based on expert opinion and not on randomized controlled studies which are the gold standard for evaluating treatment effectiveness.

Treatments for dystonia often focus on:

- Reducing pain.
- Reducing stress and anxiety.
- Increasing comfort.
- Improving sleep.
- Increasing the ability to perform activities of daily living.
- Increasing the ability to address daily personal needs independently or with a caregiver. These kinds of activities may include bathing, dressing, eating, personal hygiene, toileting and more.

Once treatment possibilities are identified, treatment should be started as soon as possible. A treatment plan can be developed with the assistance of a multi-disciplinary team that will help you choose an approach that matches your goals and what matters most to you.

Thinking Through Treatments

At this time, treatments for dystonia do not have much formal information and research on what is effective. Many medications that are prescribed for DCP are based on the research and outcomes published for people who have other types of dystonia. This fact means that most of the options used for treatment of DCP are what is referred to as “off label.” When a medication is used outside of the specifications outlined in the Federal Drug Administration approved medication packaging label, it is referred to as an “off label” use or prescription. This practice is legal and very common since there aren’t enough resources (i.e., money for research) to determine how every medication affects a particular population. When considering treatment options, here are some questions to consider:

- Do you have clear information about the movement disorders that impact the person with CP? Treatment recommendations may change if there are multiple movement disorders co-existing and affecting the individual.
- What is the impact of dystonia on pain/discomfort and can it be improved?
- Can activities of daily living improve by treating the dystonia symptoms?
- Can caregiving be made easier through interventions or positioning strategies?
- How is dystonia affecting the individual and family's quality of life?
- What are the family's and/or individual's priorities and values that may inform treatment?

Treatment Types

Medications and Neurosurgical Procedures

Several medications and types of surgeries have been used to manage the symptoms of dystonia. Here are a few examples of treatments you may hear about:

- Oral medication—You may hear about several options depending on the symptoms you are trying to address, the type of dystonia (i.e., generalized or focal dystonia) and the goals you share with your clinical team.
- Botulinum toxin—The focus of this treatment is to relax muscles by injecting botulinum toxin directly into the affected muscles.
- Intrathecal baclofen—A neurosurgical treatment where small amounts of baclofen are released slowly and directly into the spinal cord area.
- Deep Brain Stimulation (DBS)—A neurosurgical procedure where electrical impulses are used to target areas of the brain that may be generating involuntary movements associated with dystonia.

Therapeutic Strategies

Therapy consultations are often used together with oral and neurosurgical interventions.

Treatments may vary depending on a person’s symptoms and their goals for self-care and daily activities. It is important for evaluations to capture information about participation in daily life, the need for communication supports, seating and positioning adjustments and possible manual symptom relief strategies such as repositioning the individual or providing specific sensory input.

Cognitive behavioral therapy (CBT) is an approach to managing thoughts in response to circumstances and negative feelings.

Relaxation, mindfulness and breathing exercises have a limited base of evidence for dystonia but appear to be a helpful tool in lessening dystonic movements.

CO-OP is another promising cognitive and personally tailored approach but evidence is still modest for use in dystonia.





The CP community is still in need of better and more effective options to address their symptoms. Ask your medical team if you are interested in understanding the current state of research for proposed medications and/or interventions.

Priorities in Dystonia in CP Research

The CP Research Network identified 10 research themes/priorities through the **Research CP Dystonia Edition program**. This program included adults with CP, parents, therapists, physicians and researchers working together to understand and identify the greatest areas of need when it comes to DCP. We are now directing research efforts toward these 10 highlighted concerns. Research is moving forward faster in the areas of treatment, screening and diagnosis, family awareness and understanding the course of DCP over time. To view the status of research on dystonia in CP visit [our website](#) and the following section:

The first 3 priorities in DCP Research (out of 10):

- 1 Develop new treatments for individuals with dystonia in CP.
- 2 Assess rehabilitation and psychological approaches to managing dystonia.
- 3 Compare the effectiveness of pharmacological and surgical treatments for dystonia. This includes in-depth studies of side effects, impact on the person's overall function and goals.

Visit the following link to see the complete **list of 10 research priorities** identified by our community members and participating members of the clinical and research communities.

For updates on our dystonia educational and quality improvement efforts please follow us on social media and register for our webinars at cprn.org.



More Terms and Information

DYSKINETIC CEREBRAL PALSY is a term that includes three different types of involuntary movements including dystonia. Athetosis and chorea are also included and when they do occur they often present together with dystonia. When chorea and athetosis are seen together, they are referred to as choreathetosis.

Components of Dyskinetic Cerebral Palsy:

- Dystonia in CP versus dystonia outside of CP—is an important distinction. Dystonia as a stand alone movement disorder can look different and respond to different treatments compared to dystonia in CP. We need more research to explain why treatment responses differ depending on the cause of the dystonia. Dystonia in CP is often the result of an early brain injury that impacts movement, coordination and posture along with other symptoms and conditions.
- Chorea—refers to an ongoing set of random movements.
- Athetosis—a slow, continuous set of random involuntary movements that prevent a stable posture. In contrast to chorea, the same regions of the body are repeatedly involved.

Other types of Cerebral Palsy

SPASTIC CEREBRAL PALSY—The majority of individuals with cerebral palsy have the spastic form (approximately 85 percent). In spastic cerebral palsy, the individual has abnormal muscle tone and the muscles are stiff, making movement difficult.¹²

ATAXIC CEREBRAL PALSY—Between 1 to 10 percent of people with CP have the ataxic form. People with ataxic CP may have challenges with balance, depth perception and coordination. It is often characterized by wobbly or shaky purposeful movements (occurring with the intention to move), difficulty with muscles overshooting or undershooting to meet a specific target, and may also involve difficulty coordinating precise finger movements for fine motor skills such as writing or using utensils.^{10,11}

Related Terms

MUSCLE TONE—The amount of resistance or tension during rest or in response to stretching.

STATUS DYSTONICUS—The development of increasingly frequent or continuous severe episodes of generalized dystonic spasms or contractions, that may require urgent medical management/hospitalization. Status dystonicus is also referred to as dystonic storm or dystonic crisis.

Additional resources and ways to participate in research

DISCLAIMER: The information contained in the CP Research Network's Dystonia in CP Toolkit is for general information purposes only. Please consult with your medical team for specific diagnosis and treatment information related to your particular condition or medical concerns. This resource is intended to help you initiate discussions and evaluate evidence with your own professional team. Consult with your medical team about potential risks and benefits for medications, treatments, interventions, and/or programs you wish to consider using for your unique situation. The Cerebral Palsy Research Network has not validated and is not responsible for any information or services provided by third parties. You are urged to use independent judgment and request the most current references and information when considering any resource associated with the provision of services related to cerebral palsy.

For more information about dystonia research and our webinars visit [our website](#), and watch [our video](#) featuring community members sharing their experiences of DCP.

For information about dystonia publications involving the CP Research Network, visit our [Publications and Presentations](#).

Want to share your experiences with DCP or other aspects of CP? Visit our [MyCP community portal](#) to participate in research, community discussions and have further access to our educational content.

AACPDM Dystonia in CP Care Pathway The DCP Care Pathway offers clinicians comprehensive information about what is known about the assessment and treatment of DCP including the current evidence that is available.

The CP Research Network works with scientific advisors who help develop and scientifically review our educational materials. The lead scientific advisors for our Dystonia Toolkit were Bhooma Aravamuthan, MD, DPhil and Jonathan Mink, MD, PhD, FAAN. They are both pediatric neurologists and movement disorder specialists who have dedicated part of their time and research to understanding the movement disorders of cerebral palsy including dystonia. We also wish to acknowledge the contributions of Darcy Fehlings, MD, MSc, Michael Kruer, MD and Laura Gilbert, DO, MBA for laying the groundwork for organizing and growing the science around dystonia in CP.

Thank you to Fayza Jaleel for helping to develop this resource.

Thank you to all of the people who allowed us into their lives and shared their experiences with dystonia so that we could learn from them.

Special thanks to Neurocrine Biosciences for sponsoring the development of the Dystonia in CP Toolkit.



Citations and References

1. Rice J, Skuza P, Baker F, Russo R, Fehlings D. Identification and measurement of dystonia in cerebral palsy. *Dev Med Child Neurol.* 2017 Dec;59(12):1249-1255. doi: 10.1111/dmcn.13502. Epub 2017 Aug 8. PMID: 28786476.
2. Mink JW. Special concerns in defining, studying, and treating dystonia in children. *Mov Disord.* 2013 Jun 15;28(7):921-5. doi: 10.1002/mds.25548. PMID: 23893449; PMCID: PMC3806453.
3. Penner M, Xie WY, Binepal N, Switzer L, Fehlings D. Characteristics of pain in children and youth with cerebral palsy. *Pediatrics.* 2013 Aug;132(2):e407-13. doi: 10.1542/peds.2013-0224. Epub 2013 Jul 15. PMID: 23858420.
4. Sanger TD, Chen D, Fehlings DL, Hallett M, Lang AE, Mink JW, Singer HS, Alter K, Ben-Pazi H, Butler EE, Chen R, Collins A, Dayanidhi S, Forssberg H, Fowler E, Gilbert DL, Gorman SL, Gormley ME Jr, Jinnah HA, Kornblau B, Krosschell KJ, Lehman RK, MacKinnon C, Malanga CJ, Mesterman R, Michaels MB, Pearson TS, Rose J, Russman BS, Sternad D, Swoboda KJ, Valero-Cuevas F. Definition and classification of hyperkinetic movements in childhood. *Mov Disord.* 2010 Aug 15;25(11):1538-49. doi: 10.1002/mds.23088. PMID: 20589866; PMCID: PMC2929378.
5. Aravamuthan B, Pearson TS, Chintalapati K, Ueda K. Under-recognition of leg dystonia in people with cerebral palsy. *Ann Child Neurol Soc.* 2023 Jun;1(2):162-167. doi: 10.1002/cns.20018. Epub 2023 Apr 5. PMID: 38464792; PMCID: PMC10923506.
6. American Academy of Cerebral Palsy and Developmental Medicine (AACPDM) Dystonia Care Pathway Work Group. Cerebral Palsy and Dystonia. AACPDM, Milwaukee, WI, AACPDM Dystonia in CP Care Pathway <https://www.aacpdm.org/publications/care-pathways/dystonia-in-cerebral-palsy>, Accessed September 16, 2024, Updated April 2024.
7. Durkin MS, Benedict RE, Christensen D, Dubois LA, Fitzgerald RT, Kirby RS, Maenner MJ, Van Naarden Braun K, Wingate MS, Yeargin-Allsopp M. Prevalence of Cerebral Palsy among 8-Year-Old Children in 2010 and Preliminary Evidence of Trends in Its Relationship to Low Birthweight. *Paediatr Perinat Epidemiol.* 2016 Sep;30(5):496-510. doi: 10.1111/ppe.12299. Epub 2016 May 23. PMID: 27215680; PMCID: PMC5351288.
8. Gilbert LA, Fehlings DL, Gross P, Kruer MC, Kwan W, Mink JW, Shusterman M, Aravamuthan BR; Cerebral Palsy Research Network Dystonia Study Group. Top 10 Research Themes for Dystonia in Cerebral Palsy: A Community-Driven Research Agenda. *Neurology.* 2022 Aug 9;99(6):237-245. doi: 10.1212/WNL.0000000000200911. Epub 2022 Jun 17. PMID: 35715199; PMCID: PMC9442618.
9. Fehlings D, Agnew B, Gimeno H, Harvey A, Himmelmann K, Lin J-P, et al. Pharmacological and neurosurgical management of cerebral palsy and dystonia: Clinical practice guideline update. *Dev Med Child Neurol.* 2024; 66: 1133-1147.
10. Hägglund G, Hollung SJ, Ahonen M, Andersen GL, Eggertsdóttir G, Gaston MS, Jahnsen R, Jeglinsky-Kankainen I, Nordbye-Nielsen K, Tresoldi I, Alriksson-Schmidt AI. Treatment of spasticity in children and adolescents with cerebral palsy in Northern Europe: a CP-North registry study. *BMC Neurol.* 2021 Jul 12;21(1):276. doi: 10.1186/s12883-021-02289-3. PMID: 34253183; PMCID: PMC8274039.
11. Horber V, Andersen GL, Arnaud C, De La Cruz J, Dakovic I, Greitane A, Hensey O, Himmelmann K, Hollody K, Horridge K, Künzle CT, Marcelli M, Ortibus E, Papavasiliou A, Perra O, Platt MJ, Rackauskaite G, Sigurdardottir S, Troha Gergeli A, Virella D, Krägeloh-Mann I, Sellier E. Prevalence, Clinical Features, Neuroimaging, and Genetic Findings in Children With Ataxic Cerebral Palsy in Europe. *Neurology.* 2023 Dec 12;101(24):e2509-e2521. doi: 10.1212/WNL.0000000000207851. Epub 2023 Oct 19. Erratum in: *Neurology.* 2024 Jul 9;103(1):e209596. doi: 10.1212/WNL.0000000000209596. PMID: 37857495; PMCID: PMC10791054.
12. Eggink H, Kremer D, Brouwer OF, Contarino MF, van Egmond ME, Elema A, Folmer K, van Hoorn JF, van de Pol LA, Roelfsema V, Tijssen MAJ. Spasticity, dyskinesia and ataxia in cerebral palsy: Are we sure we can differentiate them? *Eur J Paediatr Neurol.* 2017 Sep;21(5):703-706. doi: 10.1016/j.ejpn.2017.04.1333. Epub 2017 May 9. PMID: 28549726.
13. Center for Disease for Control. About Cerebral Palsy. Centers for Disease Control, Atlanta, GA. Accessed September 17, 2024, updated May 14, 2024.



In addition to the noted citations, the **AACPDM Dystonia Care Pathway** has been used as a general reference throughout the DCP Toolkit.



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