"Every important aspect of spastic diplegia is addressed."

—H. KERR GRAHAM, MD, FRACS

Gillette Children's Healthcare Series

SPASTIC DIPLEGIA

Bilateral Cerebral Palsy

Second Edition

Understanding and managing the condition across the lifespan: A practical guide for families

Lily Collison, MA, MSc

Editors

Jean Stout, PT, MS

Amy Schulz, PT, NCS

Candice Johnson, OTD, OTR/L

Tom F. Novacheck, MD

GILLETTE CHILDREN'S

Praise for *Spastic Diplegia—Bilateral Cerebral Palsy*

"After the success of the first edition of Spastic Diplegia—Bilateral Cerebral Palsy, *it was time for Lily Collison and the team at Gillette to update their work. In this second edition, the text has been expanded with recent research summarized and referenced. The scope is comprehensive; the writing is clear. The journey the parents of a child with spastic diplegia embark on is frequently challenging and often frustrating due to the lack of a clear and authoritative 'user's guide.' Until the publication of this book, there was no single resource that parents could turn to for comprehensive and accurate information on a wide range of relevant topics. Here, every important aspect of spastic diplegia is addressed in a clear, conversational style that is faithful to the underlying research and science. I can think of no better guide for parents and families than this excellent text."*

H. KERR GRAHAM, Professorial Fellow, University of Melbourne; Honorary Consultant, The Royal Children's Hospital, Australia

"The second edition of Spastic Diplegia *is just as powerful if not more so than the first edition. I recommend this book to those with spastic diplegia and their families. It should also be required reading for anyone who is considering working in the CP field. This book will leave a lasting impact."*

SAMANTHA MARIE LADEMANN, Student; Adult with spastic diplegia, US

"Rarely does a book so comprehensively meet the needs of multiple stakeholders. The second edition of Spastic Diplegia—Bilateral Cerebral Palsy *does just that. It is a reliable, rigorous, and incredibly readable summary of scientific and practical information about this condition. The generosity with which Lily and Tommy share their story, interwoven with facts about treatments, assessments, and aging, left me feeling reassured that great quality, usable information is available for people living with spastic diplegia. I highly recommend this book for both families and clinicians."*

CATHY MORGAN, Principal Research Fellow and Program Lead Early Detection and Early Intervention, Cerebral Palsy Alliance Research Institute; Adjunct Associate Professor, Child and Adolescent Health, University of Sydney, Australia

"This book is an exceptional and comprehensive guide to understanding cerebral palsy in general and spastic diplegia in particular. It is a must-read for parents, caregivers, and family members of children and adults living with cerebral palsy. It covers a wide range of topics, including the causes, risk factors, diagnosis, classification, management, and treatment of the condition, in an informative and accessible manner. The inclusion of personal stories and tips from individuals and families living with spastic diplegia adds valuable insight into the lived experience of the condition. Overall, this book is an outstanding resource for families, medical professionals, and anyone seeking to gain a deeper understanding of spastic diplegia."

RACHEL THOMPSON, Co-Director, Southern Family Center for Cerebral Palsy, Rady Children's Hospital, San Diego; Associate Professor, Department of Orthopaedic Surgery, University of California, San Diego, US

"As a father to a teenager with cerebral palsy, as well as a scientist dedicated to understanding the unique health care needs facing individuals with cerebral palsy across the lifespan, I found this book to be an indispensable resource. There are very few options like this, which covers such a wide array of information that is relevant not only to spastic diplegia, but also to cerebral palsy as a general condition. I encourage any parent or caregiver of a child with cerebral palsy to consider this book as a guiding set of facts and recommendations pertaining to spastic diplegia and cerebral palsy in general."

MARK D. PETERSON, Professor, University of Michigan Medical School; Parent of a teenager with CP, US

"This book is an invaluable resource for families, empowering them with the knowledge and understanding they need to navigate a cerebral palsy diagnosis, the care, and the future. It also serves as an essential guide for health care professionals, providing comprehensive information crucial for delivering quality cerebral palsy care. As an orthopedic surgeon, I highly recommend it to both parents and medical practitioners. It should be considered mandatory reading for anyone dedicated to improving outcomes for individuals with cerebral palsy.

ENDA KELLY, Consultant Pediatric Orthopedic Surgeon, Children's Health Ireland

"Lily Collison and colleagues did an absolutely stunning job of writing this wonderful and thorough book that covers the many consequences of the complex disability of bilateral CP, and aging and living with it. The personal stories are moving and will be supportive for people with CP and their families; they also provide important insights to care providers. I hope this practical guide finds its way to parts of the world with less access to health care."

WILMA VAN DER SLOT, Rehabilitation physician and senior researcher, Rijndam Rehabilitation and Department of Rehabilitation Medicine, Erasmus MC, University Medical Center, Rotterdam, Netherlands

"This is an indispensable guidebook for navigating spastic diplegia, as it is written for families with the condition. It imparts a deep understanding of the medical science and treatment pathways, supported by comprehensive evidence-based references and information resources. The author's generous sharing of her and her son's journey, and those of many others, provides valuable sign-posting, hope, and inspiration for the reader."

JEAN AND JOHN GLYNN, parents of a son with spastic diplegia, Ireland

"This book is extremely well written. I have lived with spastic diplegia—bilateral cerebral palsy for almost 70 years, and yet I learned much from reading it. Each chapter focuses on a specific stage in the development of a person with spastic diplegic CP, from before birth to older adulthood. The author has done an excellent job of balancing the medical language and approach to addressing CP symptoms with the personal and family challenges associated with living with the condition. A particular strength of the book is the mother's ongoing narrative as her son, Tommy, progresses from his postnatal CP diagnosis through puberty, adolescence, and young adulthood, including the challenges faced at each stage and the medical procedures that were chosen to address these challenges. It definitely humanizes the clinical approach to CP interventions. This is a must-read for every member of the CP community, including individuals with CP and their family and caregivers, as well as professionals."

TED CONWAY, Research Professor, College of Engineering, Florida Atlantic University; Fellow, American Association for the Advancement of Science; Fellow, American Institute for Medical and Biological Engineering; Adult with spastic diplegia, US

"This book is truly indispensable for a deeper understanding of the medical complexity and management of bilateral spastic diplegia from infancy to teen to adult, with both reliable medical information and valuable personal perspectives. With knowledge, we can comprehend, and that allows us to make the best informed decisions at a given point in time. As a parent, I have always wanted to know what the providers know about CP, but without medical training that is impossible. But armed with this clear and concise guide, my son and I can be an important part of the care conversation. It also honors the people with cerebral palsy and their caregivers by sharing their voices and wisdom. Brava to Lily Collison and the Gillette team for bringing this second edition to fruition!

LORI POLISKI, Parent of a young adult with spastic diplegia

"This extremely useful resource for people with spastic diplegia and their families digests the current state-of-the-art care for children and adults in an organized and accessible format. The personal stories that accompany each section are poignant and informative. The book will be a great companion before and after medical visits as care decisions are being considered."

GAREY NORITZ, Director, Complex Health Care, Developmental and Behavioral Pediatrics, Nationwide Children's Hospital; Professor of Pediatrics, The Ohio State University; US

SPASTIC DIPLEGIA BILATERAL CEREBRAL PALSY

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To individuals and families whose lives are affected by these conditions, to professionals who serve our community, and to all clinicians and researchers who push the knowledge base forward, we hope the books in this Healthcare Series serve you very well.

Gillette Children's acknowledges a grant from the Cerebral Palsy Foundation for the writing of this book.

All proceeds from the books in this series at Gillette Children's go to research.

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Series Foreword

You hold in your hands one book in the Gillette Children's Healthcare Series. This series was inspired by multiple factors.

It started with Lily Collison writing the first book in the series, *Spastic Diplegia–Bilateral Cerebral Palsy*. Lily has a background in medical science and is the parent of a now adult son who has spastic diplegia. Lily was convincing at the time about the value of such a book, and with the publication of that book in 2020, Gillette Children's became one of the first children's hospitals in the world to set up its own publishing arm—Gillette Children's Healthcare Press. *Spastic Diplegia–Bilateral Cerebral Palsy* received very positive reviews from both families and professionals and achieved strong sales. Unsolicited requests came in from diverse organizations across the globe for translation rights, and feedback from families told us there was a demand for books relevant to other conditions.

We listened.

We were convinced of the value of expanding from one book into a series to reflect Gillette Children's strong commitment to worldwide education. In 2021, Lily joined the press as Program Director, and very quickly, Gillette Children's formed teams to write the Healthcare Series. The series includes, in order of publication:

- *Craniosynostosis*
- *Idiopathic Scoliosis*
- *Spastic Hemiplegia—Unilateral Cerebral Palsy*
- *Spastic Quadriplegia—Bilateral Cerebral Palsy*
- *Spastic Diplegia—Bilateral Cerebral Palsy, second edition*
- *Epilepsy*
- *Spina Bifida*
- *Osteogenesis Imperfecta*
- *Scoliosis—Congenital, Neuromuscular, Syndromic, and Other Causes*

The books address each condition detailing both the medical and human story.

Mac Keith Press, long-time publisher of books on disability and the journal *Developmental Medicine and Child Neurology*, is co-publishing this series with Gillette Children's Healthcare Press.

Families and professionals working well together is key to best management of any condition. The parent is the expert of their child while the professional is the expert of the condition. These books underscore the importance of that family and professional partnership. For each title in the series, medical professionals at Gillette Children's have led the writing, and families contributed the lived experience.

These books have been written in the United States with an international lens and citing international research. However, there isn't always strong evidence to create consensus in medicine, so others may take a different view.

We hope you find the book you hold in your hands to be of great value. We collectively strive to optimize outcomes for children, adolescents, and adults living with these childhood-acquired and largely lifelong conditions.

Dr. Tom F. Novacheck

Series Introduction

The Healthcare Series seeks to optimize outcomes for those who live with childhood-acquired physical and/or neurological conditions. The conditions addressed in this series of books are complex and often have many associated challenges. Although the books focus on the biomedical aspects of each condition, we endeavor to address each condition as holistically as possible. Since the majority of people with these conditions have them for life, the life course is addressed including transition and aging issues.

Who are these books for?

These books are written for an international audience. They are primarily written for parents of young children, but also for adolescents and adults who have the condition. They are written for members of multidisciplinary teams and researchers. Finally, they are written for others, including extended family members, teachers, and students taking courses in the fields of medicine, allied health care, and education.

A worldview

The books in the series focus on evidence-based best practice, which we acknowledge is not available everywhere. It is mostly available in high-income countries (at least in urban areas, though even there, not always), but many families live away from centers of good care.

We also acknowledge that the majority of people with disabilities live in low- and middle-income countries. Improving the lives of all those with disabilities across the globe is an important goal. Developing scalable, affordable interventions is a crucial step toward achieving this. Nonetheless, the best interventions will fail if we do not first address the social determinants of health—the economic, social, and

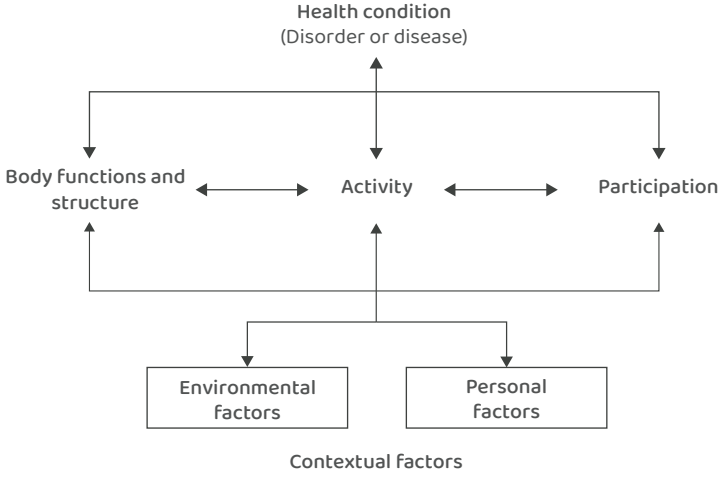
environmental conditions in which people live that shape their overall health and well-being.

No family reading these books should ever feel they have failed their child. We all struggle to do our best for our children within the limitations of our various resources and situations. Indeed, the advocacy role these books may play may help families and professionals lobby in unison for best care.

International Classification of Functioning, Disability and Health

The writing of the series of books has been informed by the International Classification of Functioning, Disability and Health (ICF).[1] The framework explains the impact of a health condition at different levels and how those levels are interconnected. It tells us to look at the full picture—to look at the person with a disability in their life situation.

The framework shows that every human being can experience a decrease in health and thereby experience some disability. It is not something that happens only to a minority of people. The ICF thus "mainstreams" disability and recognizes it as a widespread human experience.



International Classification of Functioning, Disability and Health (ICF). Reproduced with kind permission from WHO.

In health care, there has been a shift away from focusing almost exclusively on correcting issues that cause the individual's functional problems to focusing also on the individual's activity and participation. These books embrace maximizing participation for all people living with disability.

The Family

For simplicity, throughout the series we refer to "parents" and "children"; we acknowledge, however, that family structures vary. "Parent" is used as a generic term that includes grandparents, relatives, and carers (caregivers) who are raising a child. Throughout the series, we refer to male and female as the biologic sex assigned at birth. We acknowledge that this does not equate to gender identity or sexual orientation, and we respect the individuality of each person. Throughout the series we have included both "person with disability" and "disabled person," recognizing that both terms are used.

Caring for a child with a disability can be challenging and overwhelming. Having a strong social support system in place can make a difference. For the parent, balancing the needs of the child with a disability with the needs of siblings—while also meeting employment demands, nurturing a relationship with a significant other, and caring for aging parents—can sometimes feel like an enormous juggling act. Siblings may feel neglected or overlooked because of the increased attention given to the disabled child. It is crucial for parents to allocate time and resources to ensure that siblings feel valued and included in the family dynamics. Engaging siblings in the care and support of the disabled child can help foster a sense of unity and empathy within the family.

A particular challenge for a child and adolescent who has a disability, and their parent, is balancing school attendance (for both academic and social purposes) with clinical appointments and surgery. Appointments outside of school hours are encouraged. School is important because the cognitive and social abilities developed there help maximize employment opportunities when employment is a realistic goal. Indeed, technology has eliminated barriers and created opportunities that did not exist even 10 years ago.

Parents also need to find a way to prioritize self-care. Neglecting their own well-being can have detrimental effects on their mental and physical health. Think of the safety advice on an airplane: you are told that you must put on your own oxygen mask before putting on your child's. It's the same when caring for a child with a disability; parents need to take care of themselves in order to effectively care for their child *and* family. Friends, support groups, or mental health professionals can provide an outlet for parents to express their emotions, gain valuable insights, and find solace in knowing that they are not alone in their journey.

For those of you reading this book who have the condition, we hope this book gives you insights into its many nuances and complexities, acknowledges you as an expert in your own care, and provides a road map and framework for you to advocate for your needs.

Last words

This series of books seeks to be an invaluable educational resource. All proceeds from the series at Gillette Children's go to research.

CP is the most common cause of physical disability in children.[3] It is acquired during pregnancy, birth, or in early childhood, and it is a lifelong condition. There is currently no cure, nor is one imminent, but good management and treatment (addressed in Chapter 3) can help alleviate some or many of the effects of the brain injury.

When the brain injury occurs is important. The consequences of a brain injury to a fetus developing in the uterus are generally different from those of a brain injury sustained at birth, which in turn are different from those of a brain injury acquired during infancy. The European and Australian Cerebral Palsy Registers use two years of age as the cutoff for applying the diagnosis of CP.[4,5] A brain injury occurring after two years of age is called an "acquired brain injury." This two-year cutoff is applied because of the differences in brain maturity relative to when the brain injury occurs.

Although the development of movement and posture is affected in individuals with CP, as seen above, other body systems can also be affected.

How to read this book

To help you navigate the information in this book, it has been organized so that you can read it from beginning to end or, alternatively, dip into different sections and chapters independently. Because much of the information builds on previous sections and chapters, it is best to first read the book in its entirety to get an overall sense of the condition. After that, you can return to the parts that are relevant to you, knowing that you can ignore other sections or revisit them if and when they do become relevant.

This chapter addresses the overall condition of CP. Chapter 2 addresses spastic diplegia, and Chapter 3 covers the management of treatment of spastic diplegia to age 20. Chapter 4 looks at spastic diplegia in adulthood.

Throughout Chapters 1 to 4, medical information is interspersed with personal lived experience. Orange boxes are used to highlight the personal story. Chapter 5 is devoted to vignettes from individuals and families around the globe. Chapter 6 provides further reading and research.

At the back of the book, you'll find a glossary of key terms.

A companion website for this book is available at www.GilletteChildrensHealthcarePress.org. This website contains some useful web resources and appendices. A QR code to access **Useful web resources** is included below.

It may be helpful to discuss any questions you may have from reading this book with your medical professional.

My third son, Tommy, was born at term in Ireland in 1994. His older brothers, then aged four and six, had been easy babies, so by then my husband and I felt quite relaxed as parents. Tommy was born after an uneventful pregnancy and delivery, but from birth he cried incessantly. At three weeks, our family physician insisted that I give up breastfeeding. She could see how frazzled I had become due to Tommy's constant crying, his difficulty with feeding (many feeds were being returned), and the fact that neither he nor I was getting much sleep.

At three months the incessant crying suddenly stopped; Tommy became a serene, happy, and placid child, and we all relaxed again. However, a few months later, at a routine developmental check, he was deemed "developmentally delayed." That started a long journey, beginning with the diagnosis of spastic diplegia when he was a year old and continuing with the management and treatment of his condition in our community as well as at the Central Remedial Clinic* in Dublin. In the early years he also received conductive education,† and in adolescence he had a number of surgeries at Gillette Children's in Minnesota.

At the time of writing, Tommy, now 30 years old, is a college graduate, working full time, married, and living an independent life in the US.

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* Then, a national treatment center for people with physical disabilities.

† Conductive education is based on an educational rather than a medical model for treatment of children with CP. It combines educational and rehabilitation goals into a single program.

2.1

Introduction

Nothing in life is to be feared, it is only to be understood. Now is the time to understand more, so that we may fear less.

Marie Curie

The title of this book is *Spastic Diplegia—Bilateral Cerebral Palsy*. "Spastic diplegia" is the term historically used to describe this condition, and it remains in use today in the US. The term "spastic diplegia" derives from "spastic" (the type of high tone), "di" (meaning two, referring to the two affected lower limbs), and "plegia" (the Greek word for stroke). Over the past 20 years, the term "bilateral spastic CP," or simply "bilateral CP," has been adopted in Europe and Australia because it is thought to provide a more accurate description of the condition. "Bilateral" refers to two sides of the body being affected. The three terms "spastic diplegia," "bilateral spastic CP," and "bilateral CP" are all used in the scientific literature. In this book, written in the US, we use the term "spastic diplegia."

With spastic diplegia, the lower limbs are much more affected than the upper limbs, which frequently show only fine motor impairment.

Spasticity is the most common type of atypical tone present, although dystonia can be present as well.

As noted in Chapter 1, the Gross Motor Function Classification System (GMFCS) offers an indication of the severity of the condition. This book is relevant to those at GMFCS levels I, II, and III: those who are capable of walking independently or with an assistive walking device. GMFCS levels I, II, and III account for the majority of individuals with spastic diplegia.

This chapter explains spastic diplegia from birth through adolescence. It should contribute to your understanding of how the condition arises and develops over time. It provides information intended to help parents understand the diagnosis and what to anticipate as their child grows to adulthood. It provides adolescents and adults with an understanding of their condition. Chapter 3 addresses the management of the condition during childhood and adolescence. Chapter 4 is devoted to spastic diplegia in adulthood.

Physical features

Gage, an orthopedic surgeon, described the motor problems of spastic diplegia as follows:[74]

> *The involvement is primarily in the lower extremities with relatively normal upper extremity function … Most … children will walk, although balance, particularly posterior balance, is a … problem. The "typical" gait of a child with diplegia … is one of flexion, adduction, and internal rotation at the hips and flexion at the knees. The feet usually have a valgus hindfoot and … abducted forefoot.*[74]

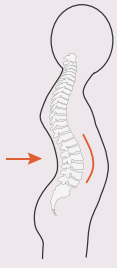
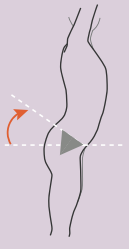
In their 2007 book, Horstmann and Bleck, also both orthopedic surgeons, described it as follows:[75]

> *When we observe the posture and gait of a child with spastic diplegia anywhere in the world it is as though each child came from the same mold. The common pattern … is flexed, adducted, and internally rotated hips … We see an increased anterior*

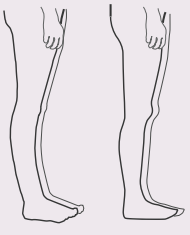
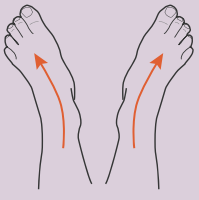
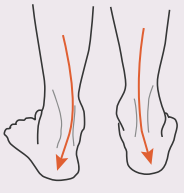
pelvic tilt, lumbar lordosis, either flexed or hyperextended knees, and equinus.

Table 2.1.1 explains and illustrates the typical physical features of spastic diplegia. Together, these features paint a picture of the movement and posture problems of spastic diplegia: of a child who walks on their toes with flexed (bent) knees and hips and whose bones are twisted.

Table 2.1.1 Typical physical features of spastic diplegia GMFCS levels I, II, and III

TERM USED IN DESCRIPTIONS	EXPLANATION	ILLUSTRATION
Lumbar lordosis	An exaggerated inward curve in the lumbar region of the spine, often called a swayback.	
Anterior pelvic tilt	A tipping forward of the pelvis to the front. (The triangle indicates the pelvis.)	
Adduction and internal rotation at the hips	Adduction is movement toward the middle of the body. Internal rotation is a twisting movement around the long axis of a bone toward the middle of the body. With adduction and internal rotation at the hips, the thigh turns inward and toward the middle of the body.	

Cont'd.

TERM USED IN DESCRIPTIONS	EXPLANATION	ILLUSTRATION
Flexion at the hips and knees	The hips and knees are bent.	
Hyperextended knees	"Hyperextended" means beyond straight or over-straightened. This is also termed "genu recurvatum." The knees on the left are hyperextended; the knees on the right are typical.	
Abducted forefoot	The front part of the foot (forefoot) moves away (outward) from the back part of the foot.	
Valgus hindfoot	The heels of both feet are turned away from the middle of the body to an atypical degree (i.e., the heels are turned outward). This is also termed "everted feet" or "eversion."	
Equinus	The toes are pointing downward (plantar flexed). In walking, this is referred to as toe walking or equinus gait.	

This chapter explains how an infant born with what appears to be typical bones, muscles, and joints can grow into a child who fits the descriptions above. I write "appears to be typical" because research is ongoing; there may be slight differences between the bones, muscles, and joints of an infant with spastic diplegia and those of a typically developing infant. Our understanding may change over time.

Distribution across classification systems

The effects of the brain injury can extend beyond movement and posture. Several classification systems for individuals with CP were introduced in section 1.7, including classification on the basis of:

- Functional mobility: Gross Motor Function Classification System (GMFCS)
- Ability to handle objects: Manual Ability Classification System (MACS)
- Communication ability: Communication Function Classification System (CFCS)
- Eating and drinking ability: Eating and Drinking Ability Classification System (EDACS)
- Visual function: Visual Function Classification System (VFCS)

Figure 2.1.1 summarizes the percentage distribution of children with spastic diplegia across the five levels of the GMFCS, MACS, CFCS, and of children with bilateral spastic CP* across the EADCS.[43–46,76–79] No data was found for the VFCS.

* Spastic diplegia and spastic quadriplegia; breakdown not available.

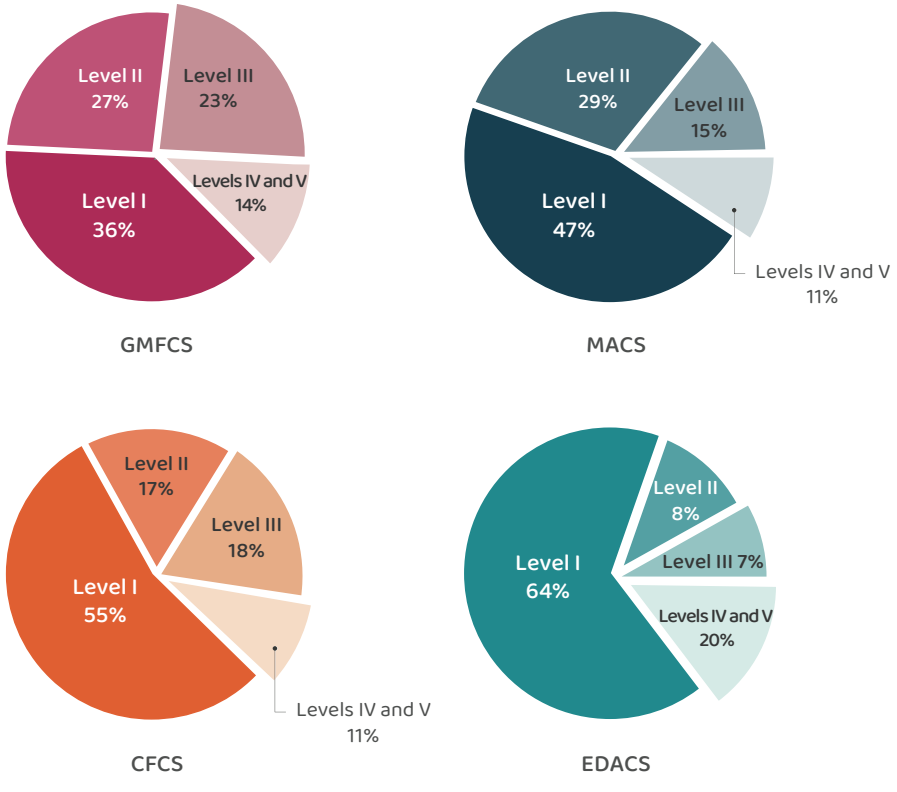


Figure 2.1.1 Distribution of children with spastic diplegia across the GMFCS,[43–46,76–78] MACS,[46,77] and CFCS[46] (top row and bottom left). Distribution of children with bilateral spastic CP across the EADCS[79] (bottom right).

Figure 2.1.1 shows that in the studies cited:

- Sixty-three percent of children with spastic diplegia were at levels I and II for GMFCS, while 76 percent were at levels I and II for MACS. This shows that the lower limbs are more affected than the upper limbs.
- Seventy-two percent of children with spastic diplegia were at levels I and II for CFCS.
- Seventy-two percent of children with bilateral spastic CP (i.e., both spastic diplegia and spastic quadriplegia) were at levels I and II for EDACS. It is expected that if those with spastic diplegia alone were considered, the percentage would be higher.

Furthermore, the level at which an individual functions on one of these classification systems can sometimes, though not always, be related to how they function on another.

Co-occurring motor type

With spastic diplegia, the predominant motor type is spasticity. However, individuals with spastic diplegia sometimes also have co-occurring, or secondary, motor types. Data from the Australian CP register shows that 12 percent of individuals with spastic diplegia (all GMFCS levels) have co-occurring dyskinesia, 2 percent have co-occurring ataxia, and 1 percent have co-occurring hypotonia.*[42] It is believed that the true prevalence of co-occurring motor types is higher[4] and the presence of dystonia with spasticity has been underrecognized.[48] A recent study found that 50 percent of children and young people with CP (all subtypes) had spasticity *and* dystonia.[48] This finding is important because the management of spasticity and dystonia is different.

Associated problems

A large Australian study reported on the prevalence of associated problems (i.e., problems with other body systems) among children aged five with spastic diplegia (all GMFCS levels).[80] See Figure 2.1.2.

* For those who acquired CP in the prenatal or perinatal period only.

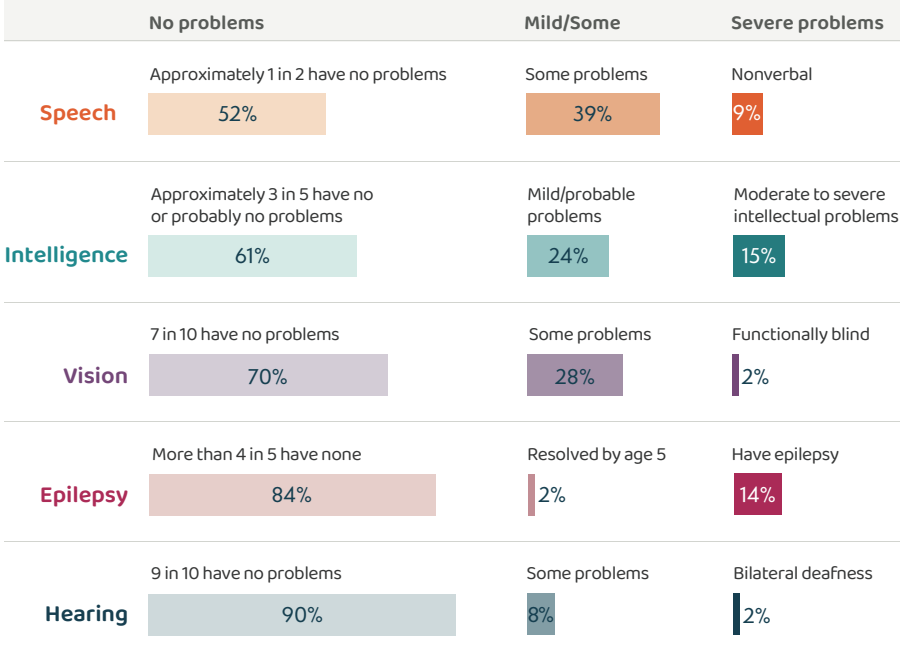


Figure 2.1.2 Prevalence of associated problems among children age five with spastic diplegia (all GMFCS levels).

Figure 2.1.2 shows that a proportion of children with spastic diplegia (all GMFCS levels) have problems in the areas of speech, intelligence (cognition), vision, epilepsy, and hearing of varying severity. Not shown in the figure is that more than 90 percent of children had none or only one severe associated problem.[80] As well, the prevalence and severity of associated problems were found to be greater in children at higher GMFCS levels compared with those at lower GMFCS levels.[80] Section 2.10 addresses associated problems in more detail.

Since spastic diplegia affects the lower limbs much more than the upper, this book focuses on the lower limbs and functional mobility for individuals with spastic diplegia.

Finally, where possible, we cite research studies relevant to those with spastic diplegia GMFCS levels I to III. Where studies include multiple subtypes, we aim to give an indication of the proportion of individuals with spastic diplegia and/or GMFCS level. Sometimes, data is available only for bilateral CP (not distinguishing diplegia and quadriplegia). We

note this where included. Sometimes, we include information about CP in general, where this is deemed useful.

USEFUL WEB RESOURCES

Tommy had a CT scan when he was approximately one year old; phrases like "significant brain damage," "not much active brain," and "go home and mind the other children" conveyed the consultant's concern about what he saw on the scan. I was perplexed: how could this consultant say these things when he had never actually met our very alert and engaging child? It turned out Tommy has normal intelligence.

Those two descriptions of physical features by Drs. Gage, Horstmann, and Bleck described Tommy pretty accurately as a young child. In the early days, I thought the way Tommy walked was particular to him. Later, as I observed other people with spastic diplegia, I realized Tommy's manner of walking was characteristic of the condition. The concept that each child with spastic diplegia "came from the same mold" really resonated with me.

In hindsight, the only differences between Tommy and his two older brothers (who do not have spastic diplegia) as a baby were that he cried incessantly for the first three months, he was difficult to feed, and his legs felt strong and stiff from birth. That early stiffness was probably spasticity, though I would only become familiar with the term much later. The photograph below shows our eldest son holding a very rigid Tommy.

Tommy and his brother, Patrick.

2.2

The brain injury

The greater danger for most of us lies not in setting our aim too high and falling short; but in setting our aim too low, and achieving our mark.

Michelangelo

In terms of brain injuries, *when* and *where* (i.e., the timing in development and the location in the brain) an injury occurs determines the effects and severity of that injury, which translates to the subtype of CP.

Spastic diplegia is most commonly associated with preterm birth or an injury in the late second or early third trimester.[74,81] In this section, we briefly address the typical brain injury that causes spastic diplegia.

The classic brain injury of spastic diplegia is periventricular leukomalacia (PVL), explained as follows:

- **Periventricular:** "Peri" means around, "ventricular" means relating to the ventricles in the brain. The ventricles* are the black areas

* Interconnected fluid-filled cavities that produce, circulate, and contain cerebrospinal fluid, which protects the brain and spinal cord.

shown in Figure 2.2.1. The injury (orange area) occurs *near* these ventricles; hence the term "periventricular," which means around the ventricles.

- **Leukomalacia:** "Leuko" means white and "malacia" means abnormal softening of tissue. The term "leukomalacia," therefore, means softening of the white tissue.

The full term, "periventricular leukomalacia," describes the injury and means softening of the white tissue (white matter) around the ventricles.

Figure 2.2.1 shows the areas of the body that may be associated with the brain injury of spastic diplegia. The white matter in the area of injury includes the motor tracts (that control movement and posture, pink lines) and sensory tracts (that deliver sensory information, green lines) between the brain and spinal cord.

The tracts closest to the ventricles relate to the ankle, those next closest relate to the knee, and those next closest relate to the hip. We will see later that this mirrors the pattern of problems in spastic diplegia: the ankle is more involved than the knee, which in turn is more involved than the hip. There is less upper limb involvement in spastic diplegia, and as shown in the diagram, those tracts are even further from the site of the injury. The injury is usually bilateral (affecting both sides of the brain), but it can be uneven. The severity of spastic diplegia depends on the timing and the magnitude of the brain injury. The injury interrupts both motor and sensory communication. Thus, while movement is affected, other body systems may also be involved.

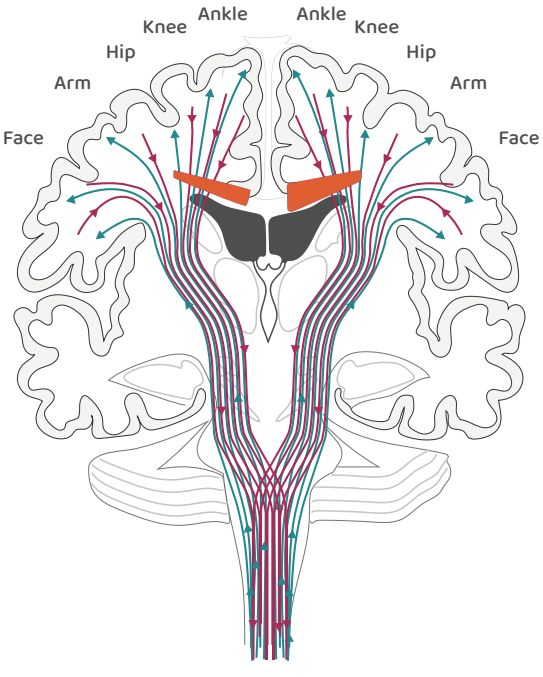


Figure 2.2.1 An example of brain injury resulting in spastic diplegia. The motor tracts (pink) descend from the cerebrum to the spinal cord, and the sensory tracts (green) ascend from the spinal cord to the cerebrum. The ventricles are the black areas. The orange area indicates the injury.

It is important to remember that this is a simplified explanation and, in reality, it is much more nuanced, unique to the individual, and complex. For example, there may be more than one area of brain injury. In addition, particularly with preterm birth, brain injury may happen more than once. The timing in development when the injury occurs is important because the areas of the brain that are developing at the time of the injury are the most vulnerable.

2.3

Growth

You have to do your own growing no matter how tall your grandfather was.
Abraham Lincoln

The musculoskeletal problems in spastic diplegia develop in proportion to growth; therefore, an understanding of growth is helpful.

Growth occurs in three major phases during a child's life: birth to age three, three years to puberty, and puberty to maturity. Of the three phases, two are of *rapid* growth: from birth to three years and during puberty.[82] The rate of growth that occurs in these two phases is particularly important for the child with spastic diplegia because musculoskeletal problems emerge with growth.

It is worth noting that there are slight differences in growth between boys and girls with CP and typically developing peers. A large US study of growth in children and adolescents with CP led to the development of growth charts for boys and girls with CP age 2 to 20. These were developed for each GMFCS level.[83,84] The study found that children and adolescents with CP (all subtypes) are shorter than typically developing

peers (see Figures 2.3.1 and 2.3.2). Similar trends in height difference among children and adolescents with CP have been observed in other parts of the world.[85]

These growth charts are included in **Useful web resources.**

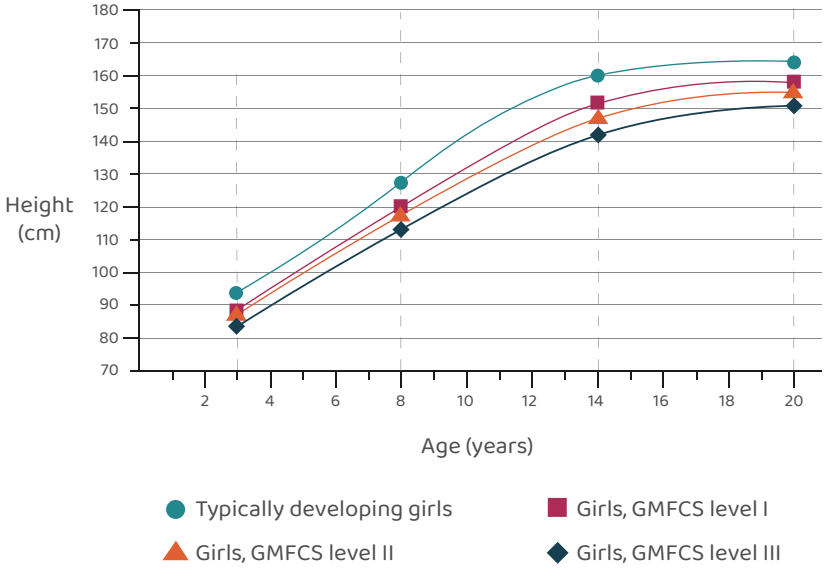


Figure 2.3.1 Height of girls with CP compared with typically developing peers. Data shows the 50th percentile height at various ages. Data collated and compiled from references.[83,84,86,87]

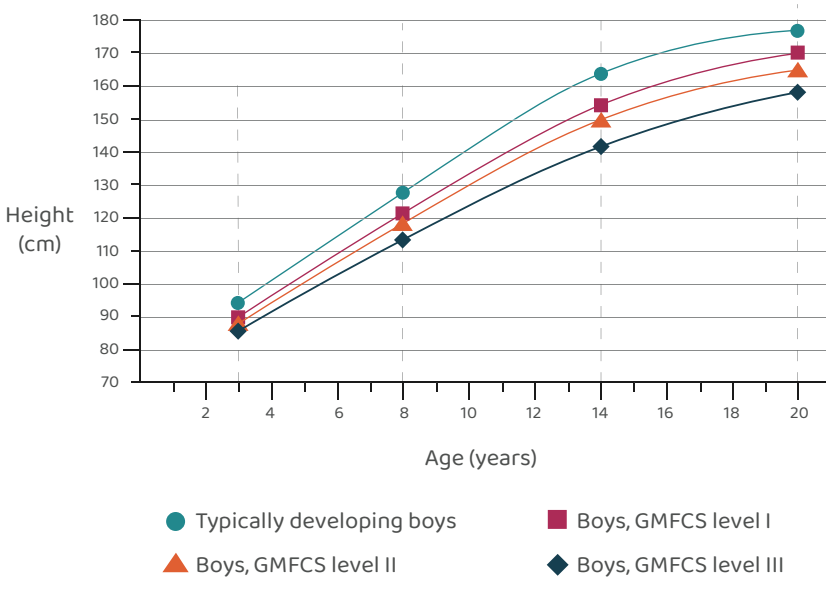


Figure 2.3.2 Height of boys with CP compared with typically developing peers. Data shows the 50th percentile height at various ages. Data collated and compiled from references.[83,84,88,89]

Since growth is such an important factor in spastic diplegia, I would advise parents to keep a growth chart for their child to stay focused on growth. I suggest you print the appropriate chart (boy or girl with CP and relevant GMFCS level) included in **Useful web links**.

Though any growth chart shows how much a typical child grows each year, an experienced parent knows that a child does not grow evenly throughout the year. Sometimes it felt like our sons shot up overnight. (I found this often followed a period of them eating more than usual.)

The difference in height between people with CP and typically developing peers was borne out at home: Tommy is not as tall as his two brothers.

The fact that the first three years are a period of very rapid growth is to some extent unfortunate as these years coincide with the time before diagnosis or when we parents are only just learning about and coming to terms with the diagnosis. This emphasizes the importance of early diagnosis so that needed early intervention can occur.

2.4

Bones, joints, muscles, and movements

It is not by muscle, speed, or physical dexterity that great things are achieved, but by reflection, force of character, and judgment.

Marcus Tullius Cicero

This section may seem like a physics and biology lesson, but because spastic diplegia affects the bones, joints, muscles, and movements, a basic understanding of them all helps enormously in understanding both the condition and its treatment.

Bones form the framework of the body, with the bones, joints, and muscles working together as levers to perform movement. In physics, a lever is a simple machine with four key components:

- A lever (a rigid bar)
- A fulcrum (a point about which the lever pivots)
- A resisting force (or load, such as a weight to be moved)
- An applied force (or effort; something that is doing the moving) (see Figure 2.4.1)

An example of a lever in humans is lifting a weight:

- The lower leg bones are the lever.
- The knee joint is the fulcrum.
- The object being lifted is the resisting force.
- The contraction of the knee extensor muscle creates the applied force.

Muscles provide the action; the bones just follow.

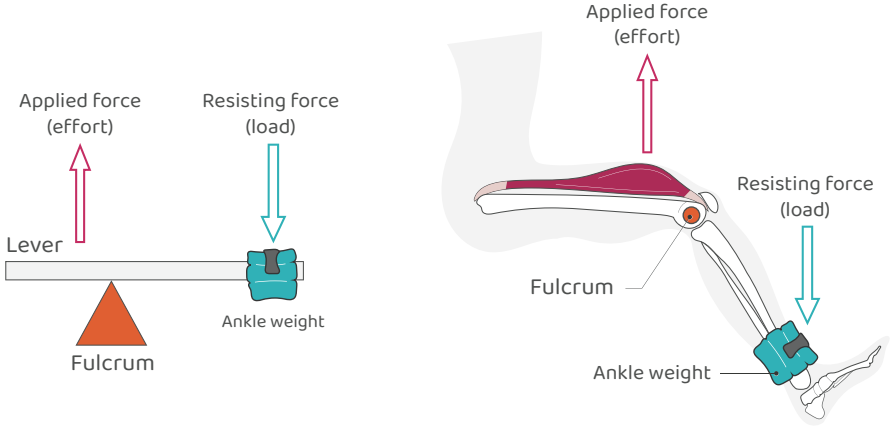


Figure 2.4.1 A lever (left) and the corresponding parts in the human leg (right).

Both the resisting force and applied force act on the lever at a distance from the fulcrum, which creates a torque or rotation (also called a "moment") about the fulcrum. This distance is called the force's "lever arm" (or "moment arm"). Even if the force stays constant, when the lever arm increases in length, the torque increases, and vice versa. Using the example in Figure 2.4.1, if the ankle weight were moved closer to the fulcrum (knee), effectively shortening the lever arm, a smaller applied force (i.e., muscle contraction) would be required to lift it.

As explained, muscles contract to produce force. The force produced can be very small (e.g., to pick up a feather) or very large (e.g., to pick up a kettlebell).

There are three types of muscles in the body:

- **Cardiac:** Muscle that forms the bulk of the wall of the heart
- **Smooth:** Muscle located in the walls of hollow internal structures such as the blood vessels, stomach, and intestines
- **Skeletal:** Muscle attached (mostly) to bones

Spastic diplegia primarily affects skeletal muscle.* Skeletal muscles contract to produce movement or maintain posture. The bones cannot stand up on their own; gravity would pull them down. When muscles contract, in addition to causing movement, they exert force, which keeps the body erect. Without these forces opposing gravity, the bones would collapse in a heap on the ground.

In a sense, the bones are like the limbs of a marionette (or puppet, see Figure 2.4.2). The marionette cannot stand up on its own.

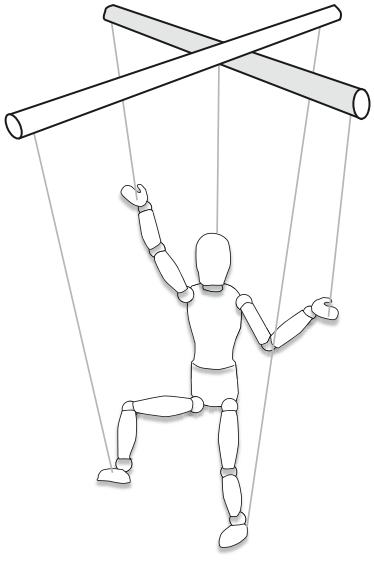


Figure 2.4.2 A marionette (puppet).

* There are some reports of smooth muscle being affected in CP.[90,91,92]

There are three types of muscle contractions:

- **Concentric (shortening):** For example, when going up a flight of stairs, the quadriceps (the muscles in front of the knee joint) contract concentrically—they shorten so that the knee extends.
- **Eccentric (lengthening):** For example, when going down a flight of stairs, the quadriceps (the same muscle that is involved in going up the stairs) contract eccentrically—they lengthen so that the knee bends. The lengthening contraction controls the bending of the knee against gravity.
- **Isometric (no change in length):** For example, when maintaining a posture (i.e., opposing the force of gravity), the muscles contract isometrically, without getting longer or shorter.

Every muscle has its own length when it is at rest. Muscles produce optimal force in the middle of that resting length.

While the details of the different types of contractions are not important in understanding spastic diplegia as such, it is helpful to keep in mind that during most movements (e.g., walking), muscles move in fractions of a second between these different types of contractions.

Muscles also contain noncontractile elements—that is, elements that are incapable of contracting. These form the tendon and various sheaths (enveloping or covering tissue). The tendon is the cord-like structure that attaches the muscle to the bone. The Achilles tendon, for example, attaches the gastrocnemius and soleus muscles—both calf muscles—to the heel. The combination of the muscle, tendon, and various sheaths is collectively known as the muscle-tendon unit (MTU).

Note also that there is a difference between muscle strength and muscle power: both are important for everyday activities like walking and running. Muscle strength is the amount of force that a muscle can generate during a specific movement—for example, the weight you can lift at the gym in a single repetition. Muscle power is the rate of force production (i.e., how fast the force is being produced). There is a strength aspect to power, but it is also about the speed of the movement. Jumping is an example of a power-based activity.

Something else to consider is range of motion (ROM), also called "range of movement," which is a measure of joint flexibility. The actual ROM through which a joint can be passively moved is measured in degrees. An instrument called a goniometer* is used to measure the ROM of a joint. (See Figure 2.4.3.) A video about measuring ROM is included in **Useful web resources.**

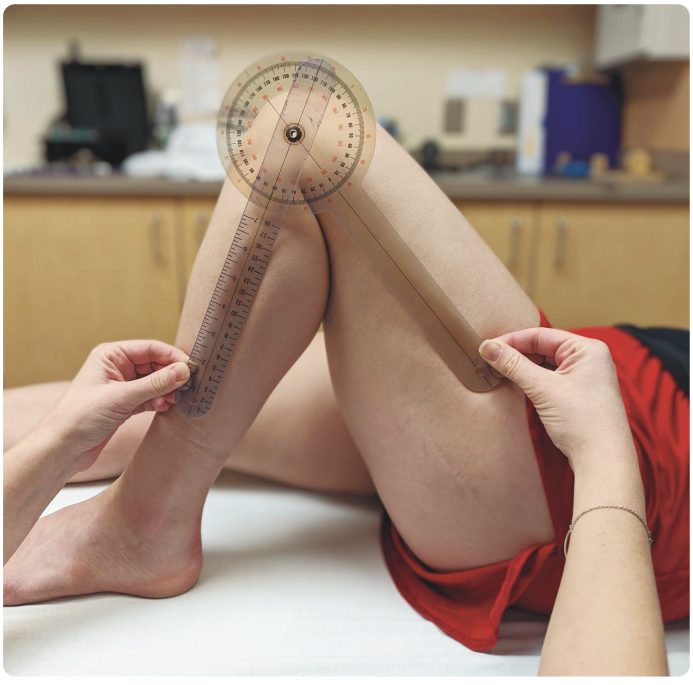


Figure 2.4.3 Measuring the ROM of the knee joint using a goniometer.

Table 2.4.1 explains the movements, joint ROMs, and key muscles for the lower limbs. This table is included as a reference and may be helpful at different times; for example, it may be useful to take it to some appointments. Below are some relevant points:

- Muscles are generally arranged in pairs around a joint. The muscles on one side of the joint move the joint in one direction, while the muscles on the other side of the joint move the joint in the opposite direction. Key muscles are identified at each joint, but minor muscles have not been included.

* A goniometer is like a movable protractor, used for measuring angles, as shown in Figure 2.4.3.

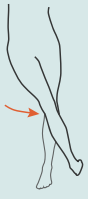
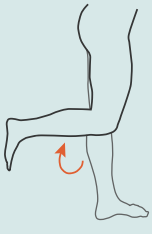
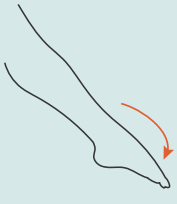
- Movements typically affected by spasticity are shown on the left side of the table and are indicated **with a green background**. *Some, but not necessarily all, of the muscles responsible for those movements may be affected by spasticity*. The table shows the movements typically affected by spasticity, but there may be some variation between individuals.
- Two-joint muscles play a role in movement at two joints. (Some muscles in the hand and foot cross more than two joints.) The most significant movements affected by two-joint muscles *and* spasticity **are indicated in orange** on the left side of the table.
- Typical ROMs for each joint are shown. The closer a joint's ROM is to typical, the better. A muscle contracture is a limitation of a joint's ROM.[93] The terms "muscle contracture" and "tight muscle" are used interchangeably in the CP field and in this book.

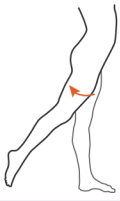
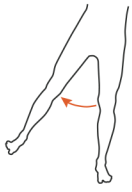
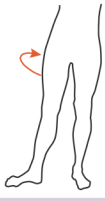
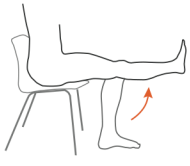
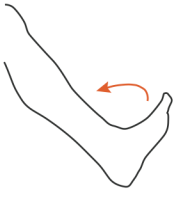
To stretch a muscle, we do the opposite of that muscle's action. To stretch a flexor muscle, for example, we must extend the joint. To stretch an extensor muscle, we must flex the joint.

To fully stretch a muscle, we must move the joint through its full ROM. Because some muscles cross two joints rather than one, both joints are involved in the stretching of two-jointed muscles. To stretch the two-jointed hamstrings, for example, we have to extend the knee while flexing the hip. Long sitting (sitting with the legs extended) is a good method of stretching the hamstrings because the knees are extended while the hips are flexed.

I compiled a version of this table many years ago, and it greatly helped my understanding of the condition. It came in handy when medical professionals referred to various movements or muscles or to a joint being tight (having a decreased ROM). I also found it helpful to be aware of the normal ROM for each joint. When surgery was planned, it helped me understand why procedures such as hip adductor and gastrocnemius lengthening were needed. For these reasons, it is worth taking some time to study this table. It will also be useful as a reference when reading later sections of this book.

Table 2.4.1 Lower limb movements, joint ROMs,[94,95] and key muscles

MOVEMENT (Green background indicates movements affected by spasticity)		KEY MUSCLES RESPONSIBLE FOR THE MOVEMENT (Two-jointed muscles are indicated in orange)
Hip flexion Movement of the thigh up toward the pelvis **ROM 0 to 125 degrees**		**Hip flexors** • Iliopsoas • Rectus femoris
Hip adduction Movement of the thigh toward the midline **ROM 0 to 20 degrees**		**Hip adductors** • Adductor longus • Adductor magnus • Adductor brevis • Gracilis
Hip internal rotation Rotary movement of the thigh toward the midline; also known as inward or medial rotation **ROM 0 to 45 degrees**		*Individual muscles not listed*
Knee flexion Increasing the angle between the thigh and lower leg **ROM 0 to 140 degrees** Note: Reference point is the straight leg. The angle increases the nearer the lower leg moves to the thigh.		**Knee flexors** • Hamstrings • Gastrocnemius
Ankle plantar flexion Movement of the foot away from the lower leg **ROM 0 to 45 degrees** Note: Reference point is the 90-degree angle between the lower leg and the foot.		**Ankle plantar flexors** • Gastrocnemius • Soleus

OPPOSITE MOVEMENT		KEY MUSCLES RESPONSIBLE FOR THE OPPOSITE MOVEMENT
Hip extension Movement of the thigh away from the pelvis **ROM 0 to 10 degrees**		**Hip extensors** • Gluteus maximus • Hamstrings
Hip abduction Movement of the thigh away from the midline **ROM 0 to 45 degrees**		**Hip abductors** • Gluteus medius
Hip external rotation Rotary movement of the thigh away from the midline; also known as outward or lateral rotation **ROM 0 to 45 degrees**		*Individual muscles not listed*
Knee extension Decreasing the angle between the thigh and lower leg **ROM 140 to 0 degrees** Note: Reference point is the flexed leg. The angle decreases the further the lower leg moves away from the thigh.		**Knee extensors** The quadriceps (quads) consist of four muscles: • Rectus femoris • Vastus intermedius • Vastus lateralis • Vastus medialis
Ankle dorsiflexion Movement of the foot toward the lower leg **ROM 0 to 20 degrees** Note: Reference point is the 90-degree angle between the lower leg and the foot.		**Ankle dorsiflexors** • Tibialis anterior • Toe extensors

2.5

Typical walking

A journey of a thousand miles begins with a single step.
Lao Tzu

In general, we take walking for granted. It is only when we encounter a problem that we stop to think about what walking entails. The term "gait" refers to a person's manner of walking. "Typical" gait refers to the typically developing person's manner of walking, which has been studied extensively. Because having problems with walking is one of the hallmarks of spastic diplegia, this section briefly looks at the features of typical walking.

Walking is a phenomenal achievement. It involves generating forces, managing gravity, speed, balance, and more. In evolutionary terms, walking on two limbs was advantageous because it freed our upper limbs for other tasks. It is no surprise that crawling comes before walking in human gross motor development: a crawling child has four limbs on the floor and is therefore more stable. Walking, which involves balancing on two limbs, is a more advanced and more demanding form of movement.

The requirements of walking

Walking has four requirements:[96]

- **A control system:** The nervous system provides the control system for walking.
- **An energy source:** The energy required is supplied by oxygen* and the breakdown of food.
- **Levers providing movement:** The levers are the bones.
- **Forces to move the levers:** Muscle contraction provides the forces for walking. As we saw in the previous section, movement is generated by muscle forces acting on the levers (the bones).

The gait cycle

One complete gait (or walking) cycle refers to the time between two successive occurrences of the same event in walking—for example, the time between when one foot strikes the ground and when that same foot strikes the ground again. Figure 2.5.1 shows what is happening with each limb during a complete gait cycle. The gait cycle is divided into two major phases:

- **Stance phase:** The period of time the foot of interest (green in Figure 2.5.1) is on the ground
- **Swing phase:** The period of time the foot of interest is in the air

Stance phase occupies approximately 60 percent of the gait cycle, and swing phase occupies approximately 40 percent.[96] There are two periods in the gait cycle when both limbs are on the ground; this is termed "double stance" (or "double support"). Single stance (or single support) is when just one limb is on the ground. Walking involves alternately balancing on each single limb as we move forward.

* Energy can be produced without oxygen in some cases; for example, for short bursts of quick walking. This is termed "anaerobic metabolism."

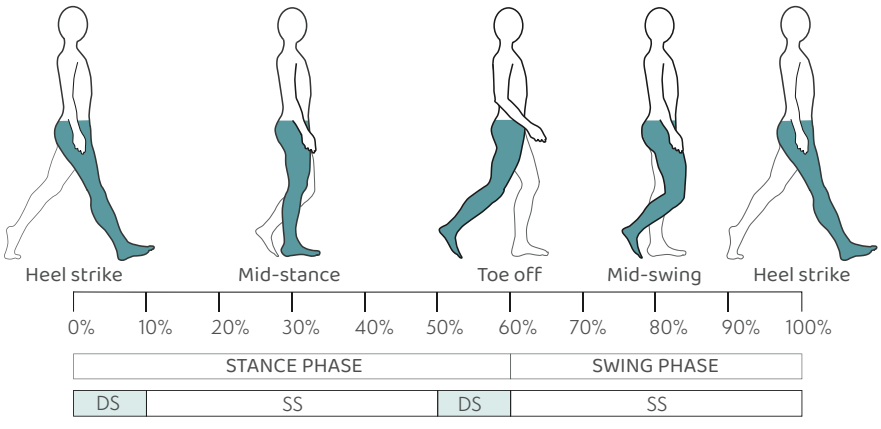


Figure 2.5.1 A complete gait cycle. "DS" is double stance; "SS" is single stance.

Attributes of typical walking

The following are attributes of typical walking that are frequently lost in individuals with spastic diplegia:[96]

- **Stability in the stance phase:** A reflection of controlled movement and good balance
- **Foot clearance in the swing phase:** Movement of the foot forward without dragging the toe
- **Pre-positioning of the foot for initial contact (heel strike):** Preparation of the foot to strike the ground with the heel (see Figure 2.5.1)
- **Adequate step length:** A sufficiently long step taken
- **Energy conservation:** Energy-efficient walking

Problems with the first four of these attributes contribute to problems with the fifth, the energy cost of walking.

When a typically developing child begins to walk, they do so without these attributes. The knees are relatively stiff and they walk with a wide base of support (i.e., the legs are far apart). But as the child develops balance and their motor system matures, their gait evolves toward the adult pattern, generally by about three and a half years of age.[97] It appears that walking is innate rather than learned, and it depends on the progressive maturing of the central nervous system.

Key points Chapter 2

- With spastic diplegia, the lower limbs are much more affected than the upper limbs, which frequently show only fine motor impairment.
- Studies show that children with spastic diplegia have more problems with gross motor function—63 percent were functioning at GMFCS levels I and II while 76 percent were functioning at MACS levels I and II.
- Spasticity is the most common type of atypical tone present in individuals with spastic diplegia, although dystonia can be present as well.
- The classic brain injury of spastic diplegia is periventricular leukomalacia (PVL). The injury is usually bilateral (affecting both sides of the brain), but it can be uneven.
- A proportion of children with spastic diplegia (all GMFCS levels) have problems in the areas of speech, intelligence (cognition), vision, epilepsy, and hearing of varying severity. However, more than 90 percent of children have none or only one severe associated problem. These and other associated problems may reduce well-being far more than motor problems.
- A useful framework for classifying the musculoskeletal problems that occur in children with spastic CP categorizes them into primary, secondary, and tertiary problems. Primary problems are caused by the brain injury and are therefore present from when the brain injury occurred. Secondary problems develop over time in the growing child. They are problems of atypical muscle growth and bone development and are referred to as "growth problems." Tertiary problems are the "coping responses" that arise to compensate for or counteract the primary and secondary problems.
- A four-group classification system describes the gait patterns in individuals with spastic diplegia.
- Crouch gait, defined as persistent flexed-knee gait, is one of the most prevalent and functional gait problems seen in spastic diplegia and varies from mild to severe.

Selective dorsal rhizotomy

Selective dorsal rhizotomy (SDR) is a neurosurgical procedure that reduces spasticity by selectively cutting abnormal sensory nerve rootlets* in the spinal cord. SDR reduces spasticity only, not other types of high tone. It is an irreversible tone-reducing treatment. The following is an explanation of each word in the full term:

- **Selective:** Only certain abnormal nerve rootlets are cut.
- **Dorsal:** "Dorsal" refers to the sensory nerve rootlets. (They are termed "dorsal" because they are located toward the back of the body. The motor nerve rootlets are termed "ventral" because they are toward the front.)
- **Rhizotomy:** "Rhizo" means "root," and "otomy" means "to cut into."

Putting it all together, "selective dorsal rhizotomy" means that certain abnormal, sensory nerve rootlets are cut to diminish the overactive reflex loop causing spasticity.

There are generally two SDR techniques, the cauda and the conus, named after the level of the spinal cord at which each procedure is performed. The choice of technique is surgeon-specific but also depends on the patient. The cauda technique is the most common worldwide.[226] Similar short-term gait and functional outcomes have been reported with each technique.[245]

SDR involves removing the back of the vertebrae (the lamina) to access the spinal cord. This is called a "laminectomy." During the operation, the sensory (dorsal) nerve roots are dissected into their individual rootlets. The rootlets are then individually electrically stimulated to determine whether they trigger a normal or abnormal (spastic) response. If a rootlet triggers an abnormal response, it is cut. If not, it is left alone. The percentage of rootlets cut varies among medical centers.[226] At Gillette Children's, the percentage of rootlets cut during SDR is lower than what is typically reported in the literature. If too high a percentage of rootlets are cut, there is greater risk of inducing weakness.

* Think of a stick of string cheese, or a telephone cable with many smaller wires together in it. The nerve root is the whole cheese stick, or the whole telephone cable, and the smaller cheese strings or wires within the cable are the rootlets.

As with many treatments, SDR is not suitable for every child. The selection criteria for SDR differs among medical centers.[226] As an example, characteristics of the "ideal" candidate for SDR at Gillette Children's include.*[246]

- Age four to seven years
- GMFCS level I–III
- Primarily spasticity (as opposed to dystonia) that interferes with function and involves multiple muscles and joints in the lower limbs
- Preterm birth history or injury in the late second or early third trimester of pregnancy
- Periventricular leukomalacia (PVL) confirmed by neuroimaging (see section 2.2)
- Energy-inefficient gait
- Satisfactory muscle strength, generally defined as antigravity muscle strength at the hips and knees
- Fair or good selective motor control at the hips and knees, meaning the child should be able to partially isolate joint movement (not moving the joint in a patterned fashion), which requires sufficient strength and motor control (i.e., the child shouldn't be reliant on spasticity for their stability or movement)
- Good ability to cooperate with rehabilitation

Children who meet all the selection criteria are uncommon, so physicians use their judgment to determine whether a particular child is a good candidate for SDR.[228] Your team will assess whether SDR is suitable for your child based on their specific circumstances.

The thinking behind the ideal age of four to seven years for SDR is that gait is relatively stable, and it is possible to decrease spasticity while the child is still young enough to learn new motor patterns. As well, secondary contracture development is usually minimal at this point, and the child is old enough to complete the assessment and cooperate with the rigorous rehabilitation program. Gait analysis is very important for evaluating whether SDR is appropriate for a particular child and for assessing outcome after surgery. (Gait analysis is addressed in the next section.)

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* Largely based on the work of Warwick Peacock, the neurosurgeon who repopularized and refined SDR in the 1980s and 1990s.

Children for whom SDR is not recommended include those with a predominance of dystonia, less severe spasticity, established contractures (typically in older children and adolescents), weakness, and poor selective motor control.[246] Though SDR is most effective in childhood, it is sometimes performed for adolescents and adults.

SDR reduces spasticity only. Any secondary problems—any muscle contractures and bone problems that are present—will remain after SDR. These may be treated by orthopedic surgery. SDR is for tone reduction; orthopedic surgery is for bone alignment and residual muscle tightness. In fact, it is important to note that as tone-reducing procedures, neither SDR nor ITB negates the need for orthopedic surgery—they are complementary procedures. The expectation should be that both tone reduction and orthopedic surgery may be needed. However, tone reduction can allow the orthopedic surgery to focus more on bone realignment than on tendon lengthening, as ROM frequently improves after SDR.

As with any treatment, prior goal-setting and being able to formally evaluate outcome is essential. SDR is a major operation, and the better the rehabilitation, the better the outcome is likely to be. Just as the operation itself varies between centers, so do post-SDR rehabilitation protocols. A review of post-SDR protocols found that patients undergo intensive PT lasting approximately one year starting in the first days after surgery. Patients remain hospitalized anywhere from six days to six weeks.[227]

As an example, at Gillette Children's, rehabilitation begins three days post-SDR and usually involves a four- to six-week hospital stay. The benefit of being an inpatient is that it allows for twice-daily therapy (PT, OT, and other therapies if necessary) and focuses on rehabilitation in this early postoperative period. Whether rehabilitation is inpatient or outpatient, the aim is to achieve the intensity of postoperative rehabilitation required to maximize functional outcomes. Information on rehabilitation after SDR at Gillette Children's is included in Appendix 6 (online).

Research supports SDR for reduced spasticity and improved gait (green light).[14] Studies of SDR showing improvements include both short- term and long-term studies (up to 26 years post-SDR).[228,247,248,249] Long-term follow-up of SDR indicates that over half the patients had orthopedic

surgery in the follow-up years.[247,248] In their long-term follow-up study (10 to 17 years after SDR), Munger and colleagues reported that non-SDR patients had significantly more orthopedic surgery and antispasticity injections than SDR patients.[249] However, a multicenter study of long-term effects of SDR found that untreated spasticity does not cause meaningful impairments in young adulthood at the level of pathophysiology,* function, or quality of life.[250]

Choosing the tone-reducing treatment

No one treatment meets every child's needs, which is why a range of tone-reducing treatments exist. Tone reduction to manage spasticity (and dystonia, if present) is tailored to the individual child's needs. Which treatment will be recommended depends on many factors, including age, GMFCS level, degree to which the tone interferes with function, and type of high tone. A child may receive different treatments as they grow. For example, they may start with BoNT-A and then undergo SDR in later childhood when their gross motor function is more mature and their ability to participate in rehabilitation has improved.[228] Though the use of tone reduction peaks in early childhood, it can continue into adolescence and adulthood.

Some tone-reducing treatments (such as SDR and ITB) are available only at specialist centers. To be able to choose the most appropriate treatment for each individual child, access to the specialist center is needed to supplement what is available locally.

Finally, note that as with all areas in the management and treatment of CP, best practice may change as more research emerges.

* The study of how a condition affects the normal functioning of the body, leading to changes in its structure or function.

While SDR is mostly carried out on young children, it can be carried out on adults. Tommy had SDR as an adult in February 2020. While it was felt that it would not achieve any major gains in terms of gait, it would be protective of his muscles in the long term given his high level of spasticity. Unfortunately, February 2020 was just before the COVID-19 pandemic, which resulted in lack of access to rehabilitation.

Tommy

SDR surgery was the first major surgery I had that wasn't on one of my legs. On the one hand, spinal surgery is more serious, given the importance of the spinal cord. But from another perspective, it was just one incision, and given that I was able to be up and about relatively quickly, the recovery seemed easier.

The first part of the rehab involved lying entirely flat for 72 hours to let the area around the spinal cord heal. The spinal cord is surrounded by fluid, and the concern is that the fluid could leak from the incision with too much movement. This 72 hours of bed rest was quite boring, but the good folks at Mssrs. Netflix, Hulu, and Apple TV provided a lot of diversion. After the enforced bed rest, you start to come up to a sitting position gradually—my memory is of something like 10 degrees per hour over an afternoon. Sensation came back slowly to my feet, and it gave rise to the very strange feeling of all 10 of my toes individually against the blanket. "Legs!" I wrote to someone at the time. "I have legs! With feet! And so many toes!" From sitting, you can return fairly quickly to normal. I took my first steps with a walker three days post-op. The big concern during this time was falling, and so almost all of my walking was done with a loop around my chest that someone behind me could keep a hold of.

Tommy and his dad.

If possible, I recommend not doing the surgery in the middle of winter, but in true glass-half-full style, I just thought of the snow and ice outside the hospital as relearning how to walk on "hard mode." Relearning to walk was trippy, in both the literal and figurative senses of the word. I would struggle to send the correct signals from brain to feet: I'd try to move my foot two inches to the left and end up six inches out in front.

I stayed on crutches for about four weeks, as long as I was an outpatient at Gillette. I think I moved onto two crutches in part to not have to transport a walker on a plane back to San Francisco. I stayed using two crutches for probably another eight or nine months. COVID impacted my ability to see a physical therapist face-to-face, but as I write this three years later, I walk independently around the house or in the office, but otherwise use a crutch for uneven surfaces, long distances, or unfamiliar terrain. Three years on, I'm glad that I chose to do SDR (although, as I said, I recommend not doing so during winter, or before a global pandemic). I'm looser, with less spasticity and less tone. This presents itself in everyday life in ways such as being able to sit cross-legged for the first time as an adult. I'm also better able to bring my knees up to my chest, such as when sitting down and putting on socks. Somewhat strangely, I lost a

lot of the ability to regulate temperature in my feet—they were very stiff and cold. I found it very uncomfortable to walk around barefoot, even just the short distance from the bedroom to the bathroom. My guess is that, since nerve endings do regulate temperature, my feet were having a harder time of this since 29 percent of those nerves were severed. With seemingly no explanation, this issue seems to have resolved itself three years on, and I can now walk around barefoot comfortably.

Key points Chapter 3

- It is important to understand what best practice in the medical care of individuals with spastic diplegia looks like. Best practice currently includes family-centered care and person-centered care, a multidisciplinary team approach, evidence-based medicine and shared decision-making, data-driven decision-making, specialist centers, early intervention, setting goals, and measurement tools and measuring outcome.
- While a lot of attention is given to development of movement and posture and secondary musculoskeletal problems, for some individuals with spastic diplegia, difficulties with communication or learning may pose bigger barriers to participation.
- Monitoring musculoskeletal development is a constant throughout childhood and adolescence.
- Hip and spine surveillance (monitoring) are important and should start early in life.
- There are two main peaks in the management of the musculoskeletal problems of spastic diplegia. The first occurs in early childhood, when tone reduction (in conjunction with other treatments such as PT and orthoses) is very important. The second occurs in later childhood (at approximately 8 to 12 years), when orthopedic surgery may be needed.
- Orthopedic surgery becomes necessary when the muscle and bone problems (the secondary problems) can no longer be adequately managed by more conservative means, and they are having a significant adverse effect on gait and function.
- Single-event multilevel surgery (SEMLS) involves multiple orthopedic surgical procedures performed during a single operation. Surgeons try to correct all the bone and muscle problems in the same surgical event to avoid multiple hospital admissions, repeated anesthesia, and multiple rehabilitations. SEMLS is now considered best practice for orthopedic surgery in CP.
- SEMLS does not alter the primary problems of CP, and a gradual recurrence of some muscle and bone problems may occur in a proportion of individuals after SEMLS.
- The home program (including exercise and physical activity) is a constant in the life of the child and adolescent with spastic diplegia.

Key points Chapter 4

- CP is diagnosed in infancy and is a lifelong condition. It is often thought of as a children's condition, but it is not. If one considers a normal life span, for every child and adolescent with CP there are approximately three adults with the condition.
- As a person with spastic diplegia reaches adulthood and skeletal growth has ceased, a certain stabilization of the musculoskeletal aspects of the condition occurs. The rate of change of the condition is slower in adulthood, assuming the adult remains physically active. People with spastic diplegia may, however, develop secondary conditions in adulthood. Some are consistent with typical aging, some may be unique. Each may influence body systems in more complex ways because of the interactions with CP itself.
- Examples of the decline that may occur with typical aging include sarcopenia, joint pain, osteoarthritis, osteoporosis, falls, and low-trauma fractures. Many conditions become more prevalent as people age, including cardiovascular disease, cancer, respiratory disease, and diabetes. These conditions are termed "noncommunicable diseases."
- Adults with spastic diplegia have had their condition since childhood, but they are also susceptible to the same challenges of typical aging. For the person with spastic diplegia, it is almost as if, on entering adulthood, two roads converge: the challenges of growing up with the condition meet the challenges of typical aging. The adult with spastic diplegia must manage these two sets of challenges in combination. The problems of aging may occur at a younger age and with more severity in adults with CP than in those without the condition.
- The prevalence of several chronic physical and mental health conditions has been found to be higher among adults with CP than those without CP. However, much can be done to prevent or minimize many of the secondary conditions that can arise.

Chapter 5

Living with spastic diplegia

Nobody gets to live life backward.
Look ahead—that's where your future lies.
Ann Landers

In this chapter, people share stories of living with spastic diplegia.

Melanie, mother of eight-year-old twins Cade and Graham, from Kentucky, US

"No wonder the heartbeat is so strong—there's TWO of them in there!"

Those were the words of the ultrasound tech one snowy morning. I had a positive pregnancy test the week before, and because I was spotting, my OB-GYN decided it would be best to see what was going on. Our first pregnancy had ended in a miscarriage; the second brought us our beautiful baby girl, Millie.

Nine-month-old Millie was sitting on my husband's lap during the ultrasound to see if our baby—BABIES—would be okay. That day set the tone for the rest of my pregnancy with our boys, Cade and Graham. After just a few months of biweekly appointments with the maternal-fetal medicine specialist, nonstop bleeding, and three trips to three different emergency rooms because I thought I was miscarrying, I went into labor at 24 weeks.

I was admitted to the hospital, and the providers were able to stop the labor. Then I went into labor again, and again it was stopped. The second time, however, my water broke, so when labor began a third time, at 25 weeks, the boys were delivered during an emergency C-section on July 6, 2015. I remember everyone in the OR being silent when both boys cried—they said at 25 weeks babies simply don't have the lung capacity to cry. My husband was able to hold the twins briefly after

they were wrapped in plastic bags and blankets and before they were whisked away to the NICU.

Everything after that was a blur.

Cade and Graham were born a couple of minutes after 10:00 on a Monday morning, and at 4:30 a.m. the next day, the NICU doctor was in my room telling me both boys had collapsed lungs and had developed pneumonia, and that he didn't expect either one to make it through the day. They did make it, but a few days later, MRIs showed they both had bilateral brain bleeds.

Over the next few weeks, it felt like the boys were competing in a race: one would show a grade II brain bleed in the next MRI, and the other would say, "I see your grade II and raise you to a grade III," until both had maxed out at grade IV bleeds on both sides.

On really bad days, I wasn't allowed to hold either of the boys. On really, really bad days, the hospital chaplain visited them. For a while, we lived minute to minute. The NICU doctors prepared us for the worst-case scenario if the boys were ever discharged from the hospital. I remember one of the nurses, after a particularly brutal conversation with a doctor about the boys' capabilities in the future, reminding me to never limit our boys, no matter what the doctors say. Eight years later, I still hold those words close to my heart. They gave me hope in that moment and continued to do so through every physical therapy session, IEP meeting, doctor's appointment, and surgery.

About a month after the twins were born, their medical issues became too much for the level 3 NICU they were in, so they were transferred to a level 4 NICU in the same city. They each had three surgeries before becoming NICU graduates on October 28, 2015, two weeks after their due date. At first we thought we were in the clear when their first post-NICU checkups showed no signs of cerebral palsy. But when days and months passed and they missed developmental milestones, it came as no surprise to get their official diagnoses when they were two years old.

In those early days, we always had people alongside us, preparing us for the time we would need to make important decisions about the boys' medical interventions. One was their physical therapist who worked

in Kentucky's Early Intervention System for children from birth to age three, who told us about different medications and procedures she had seen in her decades of experience, and offered advice on why some of them may not work for Cade and Graham, but others would. That background has allowed us to be better informed as we continue to navigate the health, education, and social systems to maximize opportunities for our boys.

To tell the stories of Cade and Graham is to also share with others their hearts. They are warriors and have had to fight to survive since day one. There has been no easy road for either of them. I couldn't tell you how many surgeries they've been through to this point, or even exactly how many diagnoses each boy has without pulling up their electronic medical records. But neither are angry or display jealousy of what other typical kids can do. Graham loves sports and remembers all the details from every event he's watched. (He may even be a little spoiled when he gets to announce batters at baseball and softball games.) Cade has a super-sweet connection to music: he can find his beat and match pitch and put all his heart into singing. Both of them are genuinely loving, trusting, resilient, and happy little boys.

So yes, that strong heartbeat the ultrasound tech heard tells the story of our boys, and there are TWO strong heartbeats. I'm so grateful that's true.

Jean, mother of nine-year-old William, from Ireland

William is an identical twin born in 2015 at 30 weeks gestation. The early birth was caused by twin-to-twin transfusion syndrome. The first diagnosis—of profound hearing loss—came within a few weeks of William's birth thanks to the national screening program that has been in place since 2011 in Ireland. In contrast, his diagnosis of cerebral palsy (spastic diplegia) wasn't made until he was almost three years old after continuing to fail his developmental milestones.

As his mother, I found it less daunting dealing with William's deafness after the initial shock of the diagnosis as I had a background in teaching

children with speech and language disorders. A number of charity groups provided immediate support and information on the treatment pathway, working in cooperation with the National Hearing Implant Centre where William had his implant done, and connected us to a welcoming and supportive network of families with a similar diagnosis. Subsequently, accessing auditory verbal therapy, which was not widely available at the time, proved critical for William to develop typical speech and language ability.

Managing William's cerebral palsy diagnosis was a very different experience. Because cerebral palsy is a one-off injury to the brain, it is essential to exploit to the maximum the brain's plasticity in childhood by early intervention. Therefore, early diagnosis and provision of comprehensive information are the basic minimums in giving every child and their families a fighting chance for best outcomes.

Unfortunately, in our case, we felt alone and isolated, with no long-term treatment pathway being offered by the health service and little information available on how to move forward. In fact, a frequent sentiment we encountered was that we should keep our expectations low for William and accept how he was rather than be informed and educated about how he could reach his full potential.

We soon realized that we had to become the explorers and managers of William's treatment, and we turned to every information source we could find to plot a way forward. Thankfully, the power of the Internet to both inform us and connect to other families with CP in Ireland and abroad, and their willingness to advise and signpost us on this journey, was the game changer.

It was through contact with parents in Ireland, the UK, and Australia that we learned about SDR surgery, which William had in the US in April 2019. After his surgery, we embarked on regular five-day intensive physiotherapy sessions in specialist centers in the UK (as none are available in Ireland), working with practitioners brought to Ireland by families to treat their children, and regularly attending intensive swimming sessions in both Spain and Ireland. Again, we found all these services through online research, group forums, and calls with parents willing to help.

William's next major milestone was SPML surgery which he had done in January 2022, again in the US. He is currently attending an orthotic center in the UK for gait support using AFOs, which he wears to school.

Today, William rides his bike and scooter, trains with the local swimming club, practices Brazilian jiujitsu, plays mini-rugby in the winter and Gaelic football and hurling in the summer, and has clawed his way to the top of the mountains in County Clare where his great-grandmother's goats once roamed. He sings, plays the tin whistle, and enjoyed earning his first "income" last year while busking with his brothers at the Fleadh Cheoil, Ireland's national traditional music festival.

William's journey is by no means over—it is just beginning—and we must continue to learn about and prepare him for the treatment pathways through adolescence and into adulthood.

Edel, mother of nine-year-old Rossa, from Ireland

My son Rossa loves the rain. I share his joy in the wildness and the relentlessness nature of driving weather, especially by the sea—the wild Atlantic waves crashing, the rhythmic ebbs and flows, furiously pushing forcefully through any boundaries in its path. It is futile to try to impress on Rossa the merits of putting up a hood, as he will laugh uncontrollably and flick it off. He embraces the magical feeling that appeals to all the senses, reminding me (and perhaps himself subliminally), that he, too, is a force of nature, unstoppable and mighty.

Following an uneventful pregnancy, my second son was born without complication. We saw many beautiful cognitive milestones in the first few months, but physical milestones and irregularities began to haunt my inner thoughts and fears. Following an MRI at 11 months, a loose diagnosis of development delay was given, which later was interpreted as cerebral palsy. Rossa had SDR at age four and a half and progressed well after the operation. Unfortunately, a seizure disorder presented when he was five and a half, and things became less predictable as we navigated through various medications.

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"A wonderful and thorough book."

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Spastic diplegia is a very common subtype of cerebral palsy (CP), and CP itself is the most common cause of childhood-onset physical disability. An estimated 17 million people worldwide have CP. Spastic diplegia is also known as bilateral spastic CP or, simply, bilateral CP.

With spastic diplegia, the lower limbs are much more affected than the upper limbs, which frequently show only fine motor impairment. Now in its second edition, this practical guide addresses spastic diplegia across the lifespan and the evidence-based, best-practice treatments. It also includes the lived experience of families.

The writing of *Spastic Diplegia* was led by Lily Collison, MA, MSc, the parent of a son with spastic diplegia. The first edition of this book gave rise to the **Gillette Children's Healthcare Series**, a series of books for families who are looking for clear, comprehensive information. Health care professionals, researchers, educators, students, and extended family members will also benefit from reading *Spastic Diplegia*.

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