"A comprehensive guide for families, health care professionals, and individuals living with this condition."

—HANK CHAMBERS, MD, PARENT

Gillette Children's Healthcare Series

SPASTIC QUADRIPLEGIA

Bilateral Cerebral Palsy

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

Marcie Ward, MD
Lily Collison, MA, MSc
Cheryl Tveit, RN, MSN, CNML
Kate Edin, Parent

Editors
Elizabeth R. Boyer, PhD
Tom F. Novacheck, MD
GILLETTE CHILDREN'S

Praise for *Spastic Quadriplegia—Bilateral Cerebral Palsy*

"Spastic Quadriplegia—Bilateral Cerebral Palsy *is a comprehensive guide designed for families, health care professionals, and individuals living with this condition. It provides a holistic approach to the medical, developmental, and emotional aspects of spastic quadriplegia. The book offers insights into cerebral palsy's causes, symptoms, and treatments, emphasizing early diagnosis, intervention, and long-term management strategies. Personal stories are interwoven throughout the text, giving a human touch to the clinical information. The experience and expertise of the team at Gillette are evident in the excellent research and writing as well as in the structure, people-centered language, and compassion in each section."*

HANK CHAMBERS, Professor of Clinical Orthopedic Surgery, University of California, San Diego; Medical Director of the Southern Family Center for Cerebral Palsy at Rady Children's Hospital, San Diego; Past President, American Academy of Cerebral Palsy and Developmental Medicine; Parent of adult son with CP, US

"Repeatedly in my reading of Spastic Quadriplegia, *I found myself wishing this book had been available 25 years ago. Parents facing a new diagnosis—or even parents who have walked this road for many years—will find the information easy to read and understand. Knowledge offers the power to prepare and to make educated decisions. Through this book, the team at Gillette Children's make that knowledge accessible to all, and thereby are certain to improve the lives of many."*

CAROL SHRADER, Mother of four, two with cerebral palsy; Policy Director for Delaware State Council for Persons with Disabilities; Freelance Writer and Speaker, US

"This book is a compelling must-read for families and nursing and medical professionals alike. It masterfully holds the space between the immense challenges of living with spastic quadriplegia and the everyday aspects of life—such as sleep, pain management, and digestion—that many take for granted but are essential to quality of life. The author reminds us to prioritize these often-overlooked needs while celebrating the joy, humour, and unique contributions that individuals with spastic quadriplegic cerebral palsy bring to the world."

IONA NOVAK, Cerebral Palsy Alliance Chair of Allied Health, The University of Sydney, Australia

"This book opens with the observation that the parent is the expert of their child while the professional is the expert of the condition. In fact, the book will go a long way to help the parent—or, indeed, the adult with spastic quadriplegia—to become an expert in the condition themselves. It will equip parents and adults with the condition to understand the bigger picture of spastic quadriplegia, whether that's by using it to come to terms with the diagnosis and preparing for a lifetime of challenges ahead, or to dip back into after being bamboozled with potential risks, therapeutic recommendations, and alternative treatment options at a medical appointment. The book methodically breaks down the latest evidence-based research into laypersons' language. It is a must-have resource for every family of a person with spastic quadriplegia."

JOHN COUGHLAN, Secretary General, International Cerebral Palsy Society; Father of a young adult with spastic quadriplegia, Luxembourg

"Spastic Quadriplegia *is a powerful and essential resource for anyone seeking to understand the complexities of this condition. With precision and compassion, it seamlessly blends cutting-edge research and clinical expertise with powerful personal narratives, creating a deeply impactful and accessible guide. What truly sets this book apart is its holistic perspective—it not only addresses the intricate medical and therapeutic challenges but also profoundly captures the strength and resilience of those living with spastic quadriplegia. This book is more than informative; it's transformative."*

RACHEL BYRNE, Executive Director, Cerebral Palsy Foundation, US

"This book presents clear and concise explanations and eliminates the confusion caused by misinformation online. It has helped me realize that we are not alone; there are other families just like ours experiencing the same highs and lows, joys and sorrows. It will provide comfort and hope to families striving to adjust to a new and oftentimes difficult diagnosis."

KRISTEN STIER, Mother of a young adult with spastic quadriplegia, US

"I highly recommend this book for parents, clinicians, and individuals affected by spastic quadriplegia. The authors provide a clear explanation of the diagnosis and treatments, empowering parents to engage in meaningful discussions with medical professionals. The journeys of individuals and families are woven throughout the text, highlighting both their struggles and their joys. This book is a must-read!"

EILEEN FOWLER, Director, Research and Education, Center for Cerebral Palsy at UCLA/ Orthopaedic Institute for Children; Past President, American Academy of Cerebral Palsy and Developmental Medicine, US

"This comprehensive guide goes beyond the clinical picture of possibilities, issues, and conditions associated with a diagnosis of spastic quadriplegia. Cowritten by a parent and with personal stories from individuals and their families with this more complex form of cerebral palsy, it offers insider insight, practical tools, and helpful advice. This book is easy to navigate and covers the lifespan; it will serve as a great starting point when new issues arise and will guide the reader to the next step in care. This insight is backed up by facts and statistics that remind us that we are not alone on this journey."

JENNIFER LYNAM, Recreational Therapist; Parent of young adult with cerebral palsy, US

"I began to read this wonderful book whilst on a flight and was happily appreciating its content and layout when I came to one of the 'orange boxes': Kate's story describing Levi's birth and perinatal trauma and the impact on both her own and her husband's life. It suddenly hit me that Kate's story was also mine. Twenty-seven years ago, I, too, had a silent placental abruption during mid–first stage of labour with my beautiful son Conor. He was born covered in blood and clots and was unresponsive. Reading the book, I relived the sadness and pain and feelings of regret that 'it' had ever happened. But I am not stuck in that time. I have moved on hugely, have survived, and have ultimately accepted Conor for the person he is today. Indeed, I very rarely think of what could/should have been. He lives in supported accommodation and is hugely loved by his dedicated carers. He is also very much loved and a vital part of our family. He visits on Sundays, and we go for walks; he tastes small amounts of chocolate, and he even goes to the pub with us while we have a drink!

"The family story told throughout the book contains wisdom and honest insights into the feelings and thoughts that accompany parents on this journey with a disabled child; they are invaluable for families and clinicians alike. It addresses what to consider when making the many care decisions, such as whether there is benefit in continuing a specific therapy if the child is not enjoying it. The photos brought the family story to life and reminded me of the many hours I spent swimming with Conor during his baby days. He loved the warmth of the water and the freedom that the buoyancy gave him.

"The book is beautifully laid out and so user-friendly; it's suitable for all levels of knowledge, and you can dip into whichever part of the book that answers your current query, whether it is in relation to feeding, growth, respiratory issues, mobility, surgery, or more. The photographs are really wonderful with so many smiling faces using the many assistive/adaptive technologies available today.

"The section on transition to adulthood raises a very important issue and is an area every country needs to work on. It is so important that parents be allowed to enjoy their late-middle and old age without having to worry about how their child will be looked after when they are no longer able to.

"Congratulations on a really wonderful book!"

DR. ÍDE NICDHONNCHA HICKEY, Principal Medical Officer, Sligo; Mother of son with spastic quadriplegia, Ireland

SPASTIC QUADRIPLEGIA 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.

Contents

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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

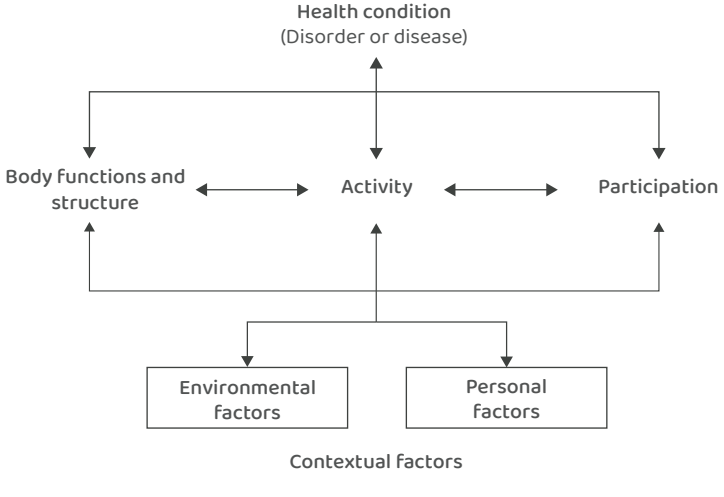
environmental conditions in which people live that shape their overall health and well-being.

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.

1.1

Introduction

So be sure when you step.
Step with care and great tact
and remember that Life's
a Great Balancing Act ...
And will you succeed?
Yes! You will, indeed!
(98 and ¾ percent guaranteed.)
Kid, you'll move mountains!

Dr. Seuss

To fully understand spastic quadriplegia, it is worth first having an understanding of the umbrella term "cerebral palsy" (CP). "Cerebral" refers to a specific part of the brain (the cerebrum) and "palsy" literally means paralysis (cerebrum paralysis). Although paralysis describes something different from the typical features of CP, it is the origin of the term "palsy."

CP was first described in 1861 by an English doctor, William Little, and for many years it was known as "Little's disease." Over the years there has been much discussion of the definition of CP, and different

definitions have been adopted and later discarded. Following is the most recently adopted definition, published in 2007:

> *Cerebral palsy describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, behaviour, by epilepsy and by secondary musculoskeletal problems.*[2]

In other words, CP is a *group* of conditions caused by an injury to the developing brain, which can result in a variety of motor and other problems that affect how the child functions. Because the injury occurs in a *developing brain and growing child*, problems often change over time, even though the brain injury itself is unchanging. Table 1.1.1 explains the terms used in the definition of CP above, in order.

Table 1.1.1 Explanation of terms in definition of CP

TERMS	EXPLANATION
Cerebral	"Cerebral" refers to the cerebrum, one of the major areas of the brain responsible for the control of movement.
Palsy	"Palsy" means paralysis, that is, an inability to activate muscles by the nervous system, though paralysis by pure definition is not a feature of CP.
Group	CP is not a single condition, unlike conditions such as type 1 diabetes. Rather, CP is a group of conditions. The location, timing, and type of brain injury vary, as do the resulting effects.
Permanent	The brain injury remains for life; CP is a permanent, lifelong condition.
Disorders	A disorder is a disruption in the usual orderly process. To meet the definition of CP, the disorder must cause activity limitation.
Posture	Posture is the way a person holds their body when, for example, standing, sitting, or moving.

Cont'd.

TERMS	EXPLANATION
Activity limitation	An activity is the execution of a task or action by an individual. Activity limitations are difficulties an individual may have in doing activities. Walking with difficulty is an example.
Nonprogressive	The brain injury does not worsen, but its effects can develop or evolve over time.
Developing fetal or infant brain	The brain of a fetus or infant has not finished developing all its neural connections and is therefore immature. An injury to an immature brain is different from an injury to a mature brain.
Motor disorders	Motor disorders are conditions affecting the ability to move and the quality of those movements.
Sensation	"Sensation" can be defined as a physical feeling or perception arising from something that happens to or that comes in contact with the body.
Perception	Perception is the ability to incorporate and interpret sensory and/or cognitive information.
Cognition	"Cognition" means the mental action or process of acquiring knowledge and understanding through thought, experience, and the senses.
Communication	Communication is the imparting or exchanging of information.
Behavior	"Behavior" refers to the way a person acts or conducts themselves.
Epilepsy	Epilepsy is a neurological disorder in which brain electrical activity becomes abnormal, causing seizures or periods of unusual behavior, sensations, and sometimes loss of awareness.
Secondary musculoskeletal problems	"Musculoskeletal" refers to both the muscles and the skeleton (i.e., the muscles, bones, joints, and their related structures). Musculoskeletal problems appear with time and growth and are therefore termed "secondary problems." They develop as a consequence of the brain injury. People with CP may develop a variety of musculoskeletal problems, such as bone torsion (twist), or muscle contracture (a limitation of range of motion of a joint).

Adapted from Rosenbaum and colleagues.[2]

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 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 quadriplegia. Chapters 3 to 6 address the overall management of spastic quadriplegia. Chapter 7 addresses alternative and complementary treatments. Chapter 8 addresses transition to adulthood, and adulthood for individuals with spastic quadriplegia is addressed in Chapter 9.

Throughout Chapters 1 to 9, medical information is interspersed with personal lived experience. Orange boxes are used to highlight the

personal story. Chapter 10 is devoted to vignettes from individuals and families around the globe. Chapter 11 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.

Final words

Before delving in, it is worth highlighting the following points, presented in no particular order:

- You may feel a mixture of feelings, some positive, some negative, and even conflicting feelings at the same time. That's okay. Sometimes you may be coping very well, other times not. That's okay too. Try to get as much support as you can. Never forget to first take care of yourself.
- You may feel incredibly overwhelmed by the sheer number of issues a diagnosis of spastic quadriplegia brings, and it can be very hard to prioritize what to tackle first, what can wait, and how to navigate everything all at once. You may feel overwhelmed that a diagnosis of spastic quadriplegia likely brings a lifetime of care; that there's a lot to organize including finding good people to help, sorting through finance and insurance issues, navigating respite, multiple appointments, and more. *We understand you and we are here to help you navigate this.*
- Early intervention is important. For example, early access to assistive technology for mobility, for communication, and for vision helps to maximize the child's ability to interact, to participate, and to learn.
- Inclusion in education early and throughout all school years is important. No child or adolescent with spastic quadriplegia should

be isolated. This will require a team approach and trialing different options to see what works best.

- Spend time exploring things that you or your child loves. Don't let therapy become the main focus or an end in itself. Many families spend so much time with therapies they don't take time to have fun and do the things they enjoy.
- There are probably far more adaptive recreational activities available to individuals with spastic quadriplegia than you think. See Chapter 4.
- Hanging out with friends and family, slowing down, and simply ensuring the individual with spastic quadriplegia is included is very important.
- Independence is a topic that you may hear a lot about, but interdependence may, in fact, be the key. In life, all of us are interdependent to varying degrees; no one is an island.
- Spastic quadriplegia is a complex condition. It is hoped that with more research, understanding will increase, and more interventions will become available. While being realistic, set your expectations high to get the best for you or your loved one with spastic quadriplegia.

USEFUL WEB RESOURCES

Levi and his identical twin brother, Cam, were born at 27 weeks gestation. Back in my 20s, I had some precancerous cells removed from my cervix through a procedure known as a cold-knife conization. Unbeknownst to me, that included removing my entire external cervix. I was lucky enough to have found out about my compromised cervix during a fertility examination prior to becoming pregnant. My OB-GYN foresaw that the removal of my cervix would make it quite difficult to carry a child to term and suggested I think about getting a cerclage (a procedure to sew up the cervix to prevent premature delivery) after I got pregnant.

In doing some research, I learned about a prepregnancy cerclage called a TAC. In this procedure, the transabdominal cerclage would be placed around the upper cervix and would perform the job of holding in a baby should I become pregnant. I flew to Chicago to have a world-renowned doctor place my TAC, and I subsequently used IUI (intrauterine insemination) to become pregnant, as my OB-GYN wanted to control as much of my pregnancy as possible.

During the cycle I got pregnant with Levi, there was only one egg follicle "ripe" enough for fertilization, so everyone was very surprised at my first ultrasound when we saw two gestational sacs! From that moment, my pregnancy went from complicated to high risk, and I switched from being attended by my regular OB-GYN to a maternal fetal medicine center. I had an irritable uterus and contracted frequently from about 14 weeks until delivery. I was on bed rest from about 20 weeks on.

At my ultrasound at week 26, my doctor released me from bed rest, but she told me to stay low-key and not play any sports. Three days later, my water broke, and in that moment the entire trajectory of my life was rerouted.

Twin A, Cameron, was the one whose water had broken. Levi was still safe and sound in his comfy bag of water. Because I had the TAC, doctors knew I would not be able to deliver naturally and would require an emergency C-section before either the babies or I got an infection. I was given a brief explanation of the neonatal intensive care unit (NICU) and handed more printouts and pamphlets about prematurity than I knew what to do with. Scared about the future, but knowing I was in the right hospital, I delivered around 9 a.m. on July 19, 2009. Cam and

Levi were both born within minutes of each other, Cam weighing 2 lb 5 oz, and Levi slightly smaller at 2 lb 2oz.

All was well for the first few hours, but then the boys began to struggle to breathe, so they both had breathing tubes inserted. In the following days, the babies did well, their brain scans were clear, and everything looked great. I began to think that everything would be okay; we would stay in the NICU until the boys were bigger, and then we would go home.

At two weeks old, Levi began to get sick. It happened quickly and was untraditional in presentation (being "untraditional" would become Levi's pattern for his entire life). His temperature spiked and his heart rate increased. He also wasn't tolerating his feeds. The doctors suspected necrotizing enterocolitis, or NEC, an infection that is prevalent in the NICU. They predicted his illness would follow the normal course and that they would have a few days to combat the illness with antibiotics.

But Levi didn't do what was expected, and within hours he was rushed into surgery because his bowels had perforated due to the NEC. The surgeon was able to remove all the dead intestine and brought the ends of his living intestine out to the surface through an ostomy (a surgically created opening). The surgery was tough, but brain scans still showed no hemorrhages or damage.

The next day, when Cam began to present with the same signs of fever and increased heart rate, doctors immediately suspected NEC and placed him on high-powered antibiotics and the JET vent (high-frequency air delivery). With that support, Cam's little body was able to fight off the infection.

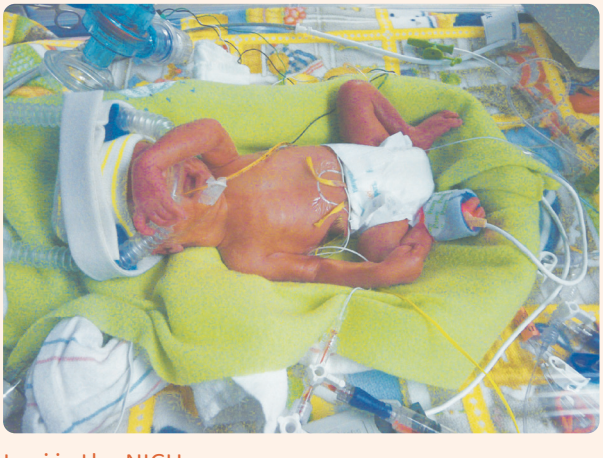
What happened next is important to understanding how Levi wound up with CP. At about eight weeks old, he was ready to have his ostomy taken down* and the ends of his intestines reconnected. The surgery was supposed to be routine, but as with everything Levi experiences, it did not go as expected. The surgeon found more necrotic tissue that needed to be removed, and Levi's little body couldn't maintain his blood

* Surgery to remove or close a previously surgically created opening (ostomy) in the abdomen. Ostomy takedown surgery is to restore bowel function by reconnecting the intestines and eliminating the need for collection of stool in an external bag.

pressure during the extensive procedure. It was after this surgery that his brain scans looked different from his brother's, and we pinpoint that as when Levi's CP came into being.

When the doctors first discussed the brain scan with me, they threw around acronyms like CP and PVL (periventricular leukomalacia, or injury to the white matter in the brain) and reminded me that no one can predict how a child with a brain scan like Levi's would do. They said they thought he would likely have ankle and foot issues based on the imaging. The white matter in Levi's brain was impacted by the PVL, but the gray matter was not, which was significant when Levi was eventually diagnosed with CP.

I went home and researched cerebral palsy and PVL. I was overwhelmed and unsure of what to do. Although I did not know it at the time, Levi's birth was also a second birth for me. I had gone from being a typical 29-year-old first-time mom of twins to being a special needs mama bear whose life focus was now learning how to support, empower, and advocate for her child.



Levi in the NICU.

Key points Chapter 1

- CP is a group of conditions caused by an injury to the developing brain that can result in a variety of motor and other problems that affect how the child functions. Because the injury occurs in a developing brain and growing child, problems often change over time, even though the brain injury itself is unchanging.
- CP is a lifelong condition. There is currently no cure, nor is one imminent, but good management and treatment can help alleviate some or many of the effects of the brain injury.
- Seventy to 80 percent of CP cases are associated with prenatal factors, and birth asphyxia (insufficient oxygen during birth) plays a relatively minor role.
- Infants who are born preterm (earlier than 37 weeks) or who have low birth weight have a higher risk of CP.
- The current CP birth prevalence for high-income countries is declining and is now 1.6 per 1,000 live births. It is higher for low- and middle-income countries.
- The most recent report from the Australian Cerebral Palsy Register shows a decrease in both prevalence and severity of CP.
- Using certain standardized tests in combination with clinical examination and medical history, a diagnosis of CP can often accurately be made before six months corrected age.
- Confirmation of the presence of a brain injury by magnetic resonance imaging (MRI) occurs in many but not all individuals with CP. Up to 17 percent of children given the diagnosis of CP have normal MRI brain scans.
- Early diagnosis is very important because it allows for early intervention. Early intervention helps to achieve better functional outcomes for the child.
- CP can be classified based on the predominant motor type (the predominant abnormal muscle tone and movement impairment) and topography (area of the body affected).
- A number of classification systems describe the functional mobility (as in movement from place to place), manual ability (ability to handle objects), communication ability, eating and drinking ability, and visual function of individuals with CP.

2.1

Introduction

You see things; and you say "Why?" But I dream things that never were; and I say "Why not?"

George Bernard Shaw

This book addresses spastic quadriplegic CP GMFCS levels IV and V. This is a complex or severe form of CP. This chapter aims to contribute to understanding how the condition arises and develops over time. It provides much of the information that parents want to know early on as they consider the future for their newly diagnosed child. Every child with spastic quadriplegia GMFCS levels IV and V is unique and has their own individual strengths and challenges.

Receiving a diagnosis of spastic quadriplegia is difficult. Children with spastic quadriplegia are often medically complex, and their care can be challenging for both the family and themselves.

The complex care needs of children with spastic quadriplegia necessitate a multidisciplinary approach, as no single profession or discipline possesses the comprehensive expertise or range to address all aspects of their care.[74] The system of care delivery for these medically complex

children is often termed the "medical home."[75] The medical home is a model for "providing accessible, family-centered, continuous, comprehensive, coordinated, compassionate, and culturally effective care to patients with the goal of improved health outcomes."[75]

Children with spastic quadriplegia are frequently born at term and may have an extensive brain injury.[3] Spastic quadriplegia involves the upper and the lower limbs and trunk; the degree of involvement often varies between the upper and the lower limbs and between the two sides of the body. The diagnosis of spastic quadriplegia is usually made early in life when the child is in a period of rapid growth. At that time, the child's joints are still flexible, but their affected muscles are already beginning to pull on their bones and joints in generally predictable patterns. This contributes to changes in posture and positioning that often can be manageable when the child is small but become more difficult as they grow. As growth continues, they may develop muscle contractures* and stiffness in their muscles and joints. Contractures occur when the muscles and tendons become tight and shortened causing flexing (bending) or stiffening of joints. This commonly occurs in the shoulders, elbows, wrists, hips, knees, and feet.

Individuals with spastic quadriplegia frequently have a distinct appearance due to muscle contractures and stiffness. Muscle contractures and stiffness can lead to functional limitations in performing daily activities, including mobility, dressing, grooming, and eating, and can often cause pain and discomfort. Some individuals with spastic quadriplegia may have difficulty smiling or frowning due to muscle and movement challenges in their face. In addition, some may have unwanted movements.

Distribution across classification systems

In Chapter 1, we saw how CP could be classified using a number of classification systems including functional mobility (GMFCS), ability to handle objects (MACS), communication ability (CFCS), eating and drinking ability (EDACS), and visual function (VFCS). Figure 2.1.1 summarizes the percentage of children with spastic quadriplegia across

* A muscle contracture is a limitation of a joint's range of motion (ROM).[76] The terms "muscle contracture" and "tight muscle" are used interchangeably in the CP field and in this book.

the five levels of the GMFCS and MACS.[45,77,78,79] No studies were found that separately identified children with spastic quadriplegia across the five levels of the CFCS, EDACS, and VFCS, but in general, though not always, they are at higher levels for those classification systems also.

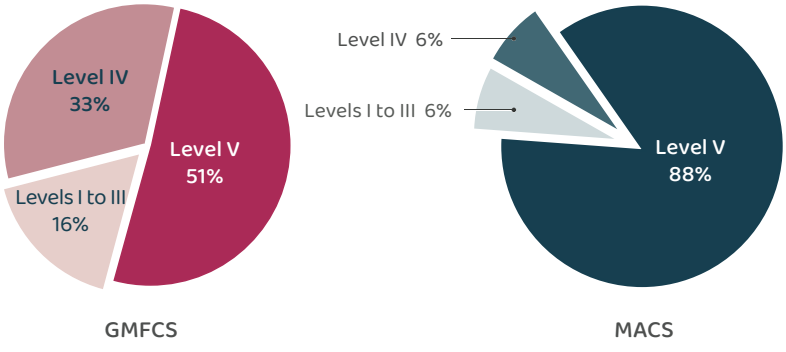


Figure 2.1.1 Distribution of children with spastic quadriplegia across the GMFCS[45,77,78,79] and MACS.[78]

Figure 2.1.1 shows that for children with spastic quadriplegia, 84 percent were at levels IV and V for functional mobility (GMFCS) and 94 percent were at levels IV and V for ability to handle objects (MACS).

This book is relevant to those at GMFCS levels IV and V, which account for the majority of individuals with spastic quadriplegia.

Co-occurring motor type

In Chapter 1, we saw that CP can be classified based on the predominant motor type, which in the case of spastic quadriplegia is spasticity. Individuals with spastic quadriplegia often have secondary, or co-occurring, motor types, hence the term "mixed movement disorder." Data from the Australian CP register shows that 48 percent of individuals with spastic quadriplegia have co-occurring dyskinesia, while 3 percent have co-occurring hypotonia.*[42] It is believed that the true prevalence of co-occurring motor types is higher.[4] A more recent study found that 50 percent of children and young people with CP (all types) had spasticity and dystonia. The presence of dystonia with spasticity

* For those who acquired CP in the prenatal or perinatal period only; also includes triplegia.

has been underrecognized.[48] This is important because the management of spasticity and dystonia is different.

Associated problems

A large Australian study reported the prevalence of associated problems among children age five with spastic quadriplegia across all GMFCS levels.[80] See Figure 2.1.2.

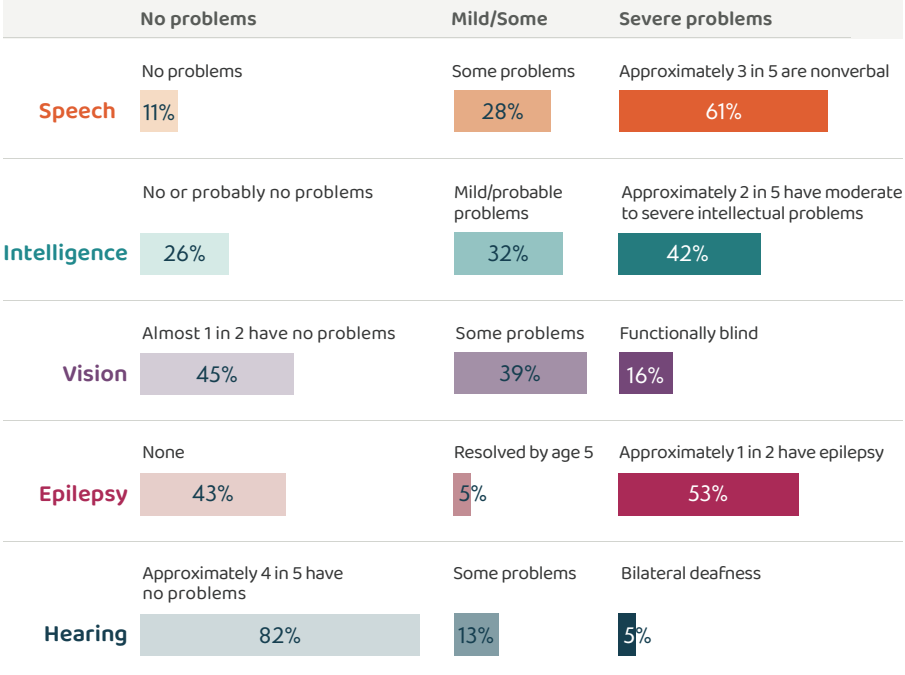


Figure 2.1.2 Prevalence of associated problems among children age five with spastic quadriplegia (all GMFCS levels). Data also includes children with triplegia.

Figure 2.1.2 shows that almost all children age five with spastic quadriplegia have problems with speech, and three in five are nonverbal. Three-quarters of children have some level of intellectual problems. More than half have some level of vision problems, and more than half have epilepsy. However, hearing is unaffected in many. Not shown in the figure is that half the children have two or more severe associated problems.[80] 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]

In this chapter, we address the brain injury and explain how it affects the development of the musculoskeletal problems. However, spastic quadriplegia affects many more body systems, and its effects may reduce well-being far more than the musculoskeletal problems. The effects of spastic quadriplegia across other body systems is addressed in Chapter 3.

Finally, where possible, the research studies cited here are relevant to those with spastic quadriplegia GMFCS levels IV and V. In those studies that include multiple subtypes, we aim to indicate the proportion of individuals with spastic quadriplegia and/or GMFCS level. Where data was available only for bilateral CP, we have included that. Sometimes, information about CP in general is included, where it is useful.

USEFUL WEB RESOURCES

2.3

Overall management philosophy

When there is no turning back, then we should concern ourselves only with the best way of going forward.

Paulo Coelho

For the child and adolescent with spastic quadriplegia, the management of all aspects of the condition can be a full-time job for parents. Some children and adolescents with spastic quadriplegia are generally quite healthy while others have many challenges. There is, as a matter of necessity, a way to prioritize management—it begins with assuring general health before addressing participation. For example, if a person is struggling with chronic respiratory or stomach issues, they do not have the energy to put toward other tasks such as development, play, or learning. This is true for all of us. Our general health is the foundation that allows us to participate in life. Therefore, this section lays out care priorities and an overall management philosophy for children and adolescents with spastic quadriplegia. And while we address management under different headings, it is crucial to bear in mind that the body operates as a unified entity, and body systems are intricately interlinked and interdependent.

Level 1 considerations, general health, are addressed in Chapter 3:

- Respiratory system
- Feeding, swallowing, and nutrition
- Digestive system
- Urinary system
- Epilepsy
- Sleep
- Pain
- Sensory system

Level 2 considerations, developmental progress and maximizing function, are addressed in Chapter 4:

- Therapies
- The home program
- Assistive technology

Level 3 considerations, musculoskeletal health–orthopedic care, are addressed in Chapter 5:

- Tone reduction
- Musculoskeletal surveillance
- Orthopedic surgery

Level 4 considerations, increasing participation, are addressed in Chapter 6:

- Cognition and intelligence
- Mental and behavioral health
- Puberty and sexual expression
- Community integration

Chapter 7 addresses alternative and complementary treatments. Chapter 8 addresses the transition to adulthood, which is then addressed in Chapter 9.

For those who would like more background reading, an explanation of the development of function is included in **Useful web resources.**

However, before we address management of spastic quadriplegia in Chapters 3 to 9, we first explain the brain injury and the musculoskeletal problems associated with the condition.

Key points Chapter 2

- Spastic quadriplegia involves the upper and the lower limbs and trunk. The degree of involvement often varies between the upper and the lower limbs and between the two sides of the body.
- The majority of children with spastic quadriplegia are at levels IV and V for functional mobility (GMFCS) and for ability to handle objects (MACS).
- The most common causes of spastic quadriplegia are hypoxic ischemic encephalopathy, periventricular leukomalacia, and central nervous system infections.
- A useful framework for classifying the neuromusculoskeletal 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.
- Spasticity is the most common type of atypical tone present in individuals with spastic quadriplegia, with 48 percent having co-occurring dyskinesia, and 3 percent having co-occurring hypotonia.
- A majority of children with spastic quadriplegia (all GMFCS levels) have associated problems varying from mild to severe in the areas of speech, intelligence (cognition), vision, and epilepsy. However, hearing is unaffected in many. Half have two or more severe associated problems.
- Prioritizing management of spastic quadriplegia begins with assuring general health, followed by addressing developmental progress and maximizing function, musculoskeletal health, and increasing participation.
- Common muscle and bone problems in spastic quadriplegia include upper limb contractures, scoliosis, hip displacement (subluxation/dislocation), windswept hips, lower limb contractures, and bone health problems.

3.2

Respiratory system

Inhale the future, exhale the past.
Anonymous

It's no surprise that healthy lung function is vital to overall health. The lungs supply oxygen to the blood so that the cells of the body can get the oxygen they need. This is why the lung is called a "vital" organ. Supplying oxygen to the body requires healthy and strong lungs. The requirements for breathing and healthy and strong lungs include:

- Head and trunk positioning to allow for easy movement of air in and out.
- An open and clear airway (which includes the nose, mouth, throat, and lungs). Any blockages in the airway can cause breathing difficulties.
- Strong respiratory muscles for breathing, but also for coughing to clear the airway.
- Sufficient surface area and functional alveoli (tiny air sacs in the lungs) to allow the exchange of oxygen and carbon dioxide.

Respiratory conditions frequently arise in individuals with spastic quadriplegia for many reasons, including the following:

- They may have difficulty clearing their airway through coughing because of their abnormal tone and muscle weakness.
- They are at high risk of **sialorrhea** (excess saliva) and **drooling** (excess saliva dropping uncontrollably from the mouth), which may then be aspirated* which can lead to respiratory problems.
- They are at high risk of aspirating generally—food, liquids, and other secretions, in addition to saliva.
- They are at an increased risk for developing asthma. At least 1 in 5 children with CP have asthma compared with 1 in 12 typically developing peers.[142]

Because of the above difficulties with airway clearance paired with higher rates of aspiration and asthma, **obstructive lung conditions**, caused by blocked airflow, can arise putting individuals at risk of recurrent lung infections and chronic inflammation.

In addition, **restrictive lung conditions**, caused by a decrease in total air volume in the lungs, can arise due to contractures and progressive scoliosis.[143] Untreated, severe, scoliosis can impact respiratory function in two distinct ways:

- The curving and rotating of the spine can limit the amount of space the lungs have to expand and fill with air, making it difficult to breathe or cough to clear the airways, especially during illness.
- Postural problems can hamper the diaphragm's ability to move the abdominal contents downwards, thus limiting the ability of the lungs to expand.

Respiratory issues are the most common cause of morbidity (temporary or permanent disability) or mortality (death) in individuals with CP.[144] Novak and colleagues reported aspiration resulting in respiratory complication as a leading cause of death in individuals with CP.[14] Thus, diligent management of the respiratory system is of utmost importance in maintaining overall health.

The good news is that there are a variety of treatments that can help to improve respiratory function and decrease the risk of respiratory illness. Having a pulmonologist as part of the care team is invaluable as

* Food or liquid entering the airway instead of the esophagus.

they can develop a comprehensive treatment plan that includes preventing problems with lung function as well as an action plan for when the lung function is becoming challenged.

- **Sialorrhea treatment:** Sialorrhea becomes a concern when the saliva is aspirated and then contributes to problems with respiratory health. When sialorrhea is causing aspiration concerns, it may help to decrease the saliva volume, which can be achieved with medications administered orally (by mouth), enterally (directly to the gastrointestinal tract through a feeding tube), buccally (placed in the cheek area), or via a transdermal patch (skin patch). Botulinum neurotoxin A (BoNT-A) injection into the salivary glands has been shown to be effective at reducing saliva.[145] If the issue is drooling, that is more a cause of social concerns and skin irritation, and for those reasons, sialorrhea may also be treated, using the same medications.
- **Suctioning:** Suctioning is a method of removing secretions to clear the airway. It can be done with a flexible catheter passed through the nose or mouth. Suctioning can sometimes cause the individual to gag or vomit, so careful attention should be given to safe positioning and gentle yet effective suction pressure levels.
- **Cough assist device:** A cough assist device typically consists of a handheld device with a face mask or mouthpiece. The device applies positive pressure to the airway, then rapidly shifts to negative pressure.* This pressure change in the airway mimics a natural cough and helps to clear secretions for individuals who have a weak natural cough.
- **Inhaled or nebulized treatments:** An inhaler or nebulizer is commonly used to administer saline solution or mucolytic medications, which help break down mucus and ease airway clearance when combined with effective coughing, suctioning, or a cough assist device.
    - An inhaler is a small, handheld device that turns liquids into a fine spray (aerosol; i.e., droplets dispersed in air) that can then be inhaled through a mouthpiece while pushing a pump. It can be paired with a spacer (a chamber that can hold medication) for gradual inhalation (breathing in).

* Positive pressure creates a pushing effect in the airways that helps to expand the lungs and move mucus or secretions toward the mouth for coughing or spitting out. Negative pressure is like a suctioning effect that helps to pull mucus or secretions from the smaller airways into the larger ones, aiding in their removal.

Key points Chapter 3

- General health considerations are addressed across individual body systems, but it is important to remember that they are all interconnected.
- Respiratory issues are the most common cause of morbidity or mortality in individuals with CP. Individuals with spastic quadriplegia face a high risk of developing chronic lung conditions. There are a variety of treatments that can help to improve respiratory function and decrease the risk of respiratory illness.
- Individuals with spastic quadriplegia often have challenges with feeding and swallowing, which can give rise to problems with nutrition. When these challenges interfere with safe swallowing or efficient mealtimes, the multidisciplinary team may recommend a feeding tube to supplement nutrition.
- Digestive challenges are common; they can cause pain, impact respiratory health, and contribute to other medical problems. Staying on top of digestive health is important for the overall health and quality of life of the individual with spastic quadriplegia.
- While incontinence is the most common urinary system problem, other problems include the need to urinate frequently and urgently, hesitancy, and urinary retention.
- Epilepsy is common among individuals with spastic quadriplegia. Treatments are available.
- Adequate sleep is necessary for brain development, and the disruption of sleep and sleep deprivation has been shown to have many negative effects. Sleep medicine specialists can help to evaluate sleep problems and identify solutions.
- Pain is common among individuals with spastic quadriplegia who may not always be able to communicate that they are in pain. Involving specialists in pain and palliative care, medical professionals who focus on alleviating pain with a multimodal approach, should be considered.
- Sensory problems can include problems with vision and processing visual input, hearing and processing auditory input, and decreased sensation and a lack of awareness of the position and movement of the body (proprioception).
- Addressing general health issues as comprehensively as possible serves as a foundation allowing individuals to more fully participate in life.

4.4

Assistive technology

Determine that the thing can and shall be done,
and then we shall find the way.
Abraham Lincoln

"Assistive technology"* refers to products and services designed to enhance the functional capabilities and independence of individuals with disabilities to allow them to participate.[195] This section addresses the following assistive technology commonly used by individuals with spastic quadriplegia:

- **Seating, standing, and mobility aids**
- **Adaptive beds and sleep systems**
- **Adaptive equipment for activities of daily living**
- **Augmentative and alternative communication**
- **Environmental control devices**
- **Orthoses**
- **Adaptive recreational equipment**

* In the US, some assistive technology is referred to as "durable medical equipment" (DME) for medical insurance purposes.

It is important to note that this is not an exhaustive list, and that assistive technology continues to improve and develop.

Assistive technology should be selected based on assessments performed by a multidisciplinary team including professionals (e.g., physician, physical and occupational therapists, speech-language pathologist, orthotist) in conjunction with the individual and their family. An individual's unique health needs and the family's care and function goals help guide what assistive technology is best suited for the individual. The family knows the individual best and their own particular home and circumstances. In some cases, it is worth doing some reading to be as informed as possible to help choose the most appropriate assistive technology. Regular reevaluation is important, the frequency of which depends on the individual and the product.

Some pointers on making your home more accessible are included in **Useful web resources**.

Seating, standing, and mobility aids

Seating, standing, and mobility aids provide improved function during sitting, standing, and movement. Individuals with spastic quadriplegia GMFCS levels IV and V often have high equipment needs for these functions. The following are commonly used by individuals with spastic quadriplegia:

a) **Floor sitters**
b) **Adaptive chairs**
c) **Standers**
d) **Gait trainers**
e) **Adaptive strollers**
f) **Wheelchairs**
g) **Transfer aids**
h) **Vehicle transportation**

a) Floor sitters

Young children do a lot of playing and learning at floor level. Floor sitters provide postural support for the child with spastic quadriplegia

Adaptive recreational equipment

A variety of adaptive recreational equipment is available to make participation in various recreational or leisure pursuits possible or just even easier. An appointment with a recreational therapist, physical therapist, or occupational therapist is useful if people need guidance. Examples of such equipment follow here:

a) Cycles
b) Adaptive workout equipment
c) Hiking aids
d) Other outdoor and adventure sports equipment
e) Technology options
f) Adaptive art and crafts equipment
g) Reading options
h) Adaptive equipment for games

a) Cycles

Adaptive cycles come in many configurations, but for the individual with quadriplegia, an adaptive tricycle allows for more stability to accommodate for balance challenges. Consideration of the transfer height to the cycle seat and the center of gravity while riding may be factors in choosing an upright style versus a recumbent* style. Other factors affecting choice include the age, size, and abilities of the cyclist. Features such as foot plates (also known as shoe holders) with straps, trunk positioning options (such as lateral pads, lap belt, and chest harness), or headrests allow for optimal safety and function. Additionally, a caregiver steering column allows for safety and control by a caregiver assisting with the steering and braking. Integration into family activities is important, so for longer rides or situations where pedaling is less important than inclusion, other cycle styles are available for the rider to simply enjoy the ride. These cycles or trailers may attach to a standard adult cycle, or there are tandem options where the wheelchair can be toted along too. See Figures 4.4.9 and 4.4.10.

* Where the cyclist is seated in a reclined or lying-back position.

Figure 4.4.9 Foot-powered adaptive tricycles.

Figure 4.4.10 A non-pedaling rider tandem adaptive cycle.

b) Adaptive workout equipment

Weight-lifting exercises may be done safely with adaptive equipment such as a wrist cuff grip to hold a free weight. They can also be done with a regular wrist weight that goes around the wrist or by using resistance bands. See Figure 4.4.11.

Figure 4.4.11 Wrist cuff grip to hold a free weight (left), wrist weight (middle), and resistance bands (right).

c) Hiking aids

There are some wearable riding options with higher weight capacities to allow a larger child or a smaller adult to ride on a caregiver's back to enjoy the outdoors.

Another option is a pull-behind chariot (a seat on wheels, attached to poles). This is also known as a tandem hiker. See Figure 4.4.12.

Figure 4.4.12 A pull-behind chariot. Reproduced with kind permission from Huckleberry Hiking.

A track chair is an all-terrain wheelchair that can safely navigate outdoor spaces. Some large parks and recreational facilities have these chairs available to borrow while at the park or at the beach, for example. See Figure 4.4.13.

Figure 4.4.13 Track chairs.

d) Other outdoor and adventure sports equipment

Adaptive equipment is available for many other outdoor and adventure sports, including:

- Swimming aids (e.g., an adaptive life jacket with extra head support for floating and/or swimming)
- Jogging strollers
- Frame running (also known as race running), where individuals compete with running frames on an athletics track
- Snow skiing (e.g., outriggers)
- Waterskiing (e.g., arm sling handles, shoulder wraps, sit skis)
- Golf (e.g., adaptive club grips and golf carts)
- Kayaking (e.g., angled oars, outriggers, trolling motors)
- Surfing (e.g., specialized systems)
- Horseback riding (e.g., high back saddles for balance)
- Adaptive baseball (e.g., use of a wrist band to hold the bat)

These can have various levels of assistance and support. See Figures 4.4.14 to 4.4.18.

Figure 4.4.14 Adaptive swimming.

Figure 4.4.15 Adaptive snow skiing.

Figure 4.4.16 Adaptive waterskiing.

Key points Chapter 4

- Physical therapy, occupational therapy, and/or speech and language pathology (or therapy) are important tools in promoting development and maximizing function for the individual with spastic quadriplegia.
- Physical therapists help the individual to maximize their movement and functional ability.
- Occupational therapists help the individual to complete meaningful tasks and help modify activities or the environment to enable successful task completion.
- Speech and language pathologists/therapists support those with speech, language, and communication needs as well as feeding and swallowing difficulties.
- "It takes a village to raise a child" emphasizes the idea that raising a child, particularly a child with a disability, is not a task that should be shouldered alone by parents. Enlisting the support of others is important. Practicing what is learned in therapy, postural management, and exercise and physical activity are important elements of the home program.
- There is a wide variety of assistive technology that can help the functional capability and independence of individuals with spastic quadriplegia. Therapists and orthotists, together with other members of the multidisciplinary team, work with the individual in this area. Assistive technology includes:
    - Seating, standing, and mobility aids to help improve function during sitting, standing, and movement; these include floor sitters, adaptive chairs, standers, gait trainers, adaptive strollers, wheelchairs, transfer aids, and vehicle transportation
    - Adaptive beds and sleep systems to enhance comfort, positioning, safety, and care
    - Adaptive equipment for activities of daily living (ADLs) to help with everyday tasks, such as adaptive bathroom equipment; eating, drinking, and dressing aids; and digital accessibility features and tools
    - Augmentative and alternative communication (AAC) to supplement or compensate for problems with expressive communication

- Environmental control devices to allow for independent control of the environment
- Orthoses to hold specific body parts in position in order to modify their structure and/or function, including lower and upper extremity and spinal orthoses
- Adaptive recreational equipment to make participation in a wide range of hobbies and sports possible or just even easier

Key points Chapter 5

- The overall goal of managing musculoskeletal health (i.e., orthopedic care) is to prevent and treat musculoskeletal problems that may interfere with function, ease of daily cares, and comfort now or in the future to allow the individual to participate as fully as possible in life.
- Managing musculoskeletal health includes tone reduction, musculoskeletal surveillance, and orthopedic surgery.
- For musculoskeletal treatments, having a multidisciplinary team and shared goal-setting between the individual with their family and medical professionals is very important, with everyone considering and weighing the benefits and risks of the intervention against those of not proceeding with the surgery.
- Treatment of high tone includes the daily home stretching program, casting, and/or orthotics, oral or enteral medications, injectable medications (BoNT-A and phenol), and neurosurgery (including intrathecal baclofen, ventral dorsal rhizotomy, and deep brain stimulation).
- Musculoskeletal surveillance involves regular monitoring and assessment of musculoskeletal development and function. It includes surveillance of lower and upper limb range of motion, hip displacement, spinal curvature, and bone health.
- Orthopedic surgery includes surgery on the lower limb, upper limb, hip, and spine.
- Hip surgery can be classified as preventive, reconstructive, and salvage. The need for salvage surgery can be avoided by early hip surveillance and with appropriately timed preventive or reconstructive surgery. Salvage surgery outcomes are not as good as interventions at earlier stages.
- Both neurosurgery and orthopedic surgery are big undertakings for individuals with spastic quadriplegia as their general health status is often more difficult to maintain and the risk of complication is higher. Preoperative general health status must be assessed and optimized and there must be a focus on excellent postoperative care.

Key points Chapter 6

- Many individuals with spastic quadriplegia GMFCS levels IV and V have intellectual challenges ranging from mild to severe.
- The diagnosis of intellectual disability is typically made during childhood or adolescence when problems in intellectual functioning and adaptive functioning become apparent.
- Neuropsychological testing helps to evaluate broad areas of cognitive function; for example, intelligence, language, visuospatial function, executive function, attention, memory, and processing speed.
- Early screening for cognitive functioning and identification of intellectual disability can help guide educational planning and supportive intervention services for the child to maximize their cognitive function. Appropriate supports (e.g., adapted education, therapy services, and assistive technology) frequently need to be put in place to maximize the educational growth of the child.
- It can be challenging to accurately measure cognitive function in children with severe motor, visual, and/or communication problems. Testing may need to be modified as appropriate and results interpreted carefully.
- Children with spastic quadriplegia are at risk for mental health problems. In nonverbal individuals, behavioral outbursts can be due to pain, hunger, fatigue, or change in routine, among other causes. Ruling out physical reasons for behaviors is an appropriate first step in behavioral management.
- Evaluations for mental and behavioral health should be incorporated into multidisciplinary team assessments and appropriate treatments prescribed.
- The onset of menstruation can present challenges for females with spastic quadriplegia, including hygiene and pain.
- Adolescents with spastic quadriplegia have the right to sexual expression and should receive appropriate support and education in this area from a multidisciplinary team, including health care providers, psychologists, and counselors.
- Maximizing activity and participation requires a comprehensive management approach. By addressing challenges, individuals with spastic quadriplegia can be supported to actively participate in society, promoting their overall well-being and quality of life.

- Strategies for community integration can include accessible transportation, inclusive recreational activities, educational accommodations, and promoting peer relationships.

Key points Chapter 9

- CP is diagnosed in childhood and is a lifelong condition. The severity of problems affects life expectancy. However, over the past three decades, data from California has shown that there have been significant improvements in the survival of children with CP and severe disability.
- Many, but not all, individuals with spastic quadriplegia will likely need supported living arrangements and support with decision-making.
- As a person with spastic quadriplegia 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. Individuals with spastic quadriplegia 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 quadriplegia have had their condition since childhood, but they are also susceptible to the same challenges of typical aging. For the person with spastic quadriplegia, 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 two sets of challenges must be managed 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 was 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 10

Living with spastic quadriplegia

What lies behind us and what lies before us
are tiny matters compared to what lies within us.
Oliver Wendell Holmes

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

Jarett, father of five-year old Julia, from Minnesota, US

It was November of 2018. In the span of a week, my wife, Erin, was diagnosed with a deadly brain cancer (glioblastoma), my father had a significant stroke, and our younger daughter was diagnosed with microcephaly (and later cerebral palsy). These were such significant events and moments for our entire family, and each one needed its own attention to detail and care, and each demanded such emotional and general processing of the immense ripple effects on our existing life plans, thoughts, and dreams. Everything changed about what we thought the next week, month, and year would and could hold for us.

We received the diagnosis for Julia in a coldly worded letter, clearly stating, "Julia has cerebral palsy." This communication approach was much different from our in-person discussion we had had with the doctors about what might be going on. We had been tracking Julia's progress because we had concerns. She was small, just like her two-and-a-half-year-old sister had been at the same age, but Julia's head size was tracking the very bottom of growth chart, she was not making physical gains like her older sister did, and she was not hitting the typical milestones. We could also see a difference in the way she was progressing compared to her peers at daycare.

After receiving the letter, my wife and I wanted to know the "why": Why does our daughter have cerebral palsy? How did this happen? We embarked on some genetic testing for answers, but it took us nowhere.

Erin was experiencing major "mom guilt," as she felt like she had done something wrong, and what was happening to Julia was her fault in some way. Making it extra difficult was she was processing these feelings alongside her own terminal brain cancer diagnosis. I reminded her of all the times she did everything she could, how she did things right, how she took care of herself and Julia during her pregnancy, how much she loved and cared about Julia—and that none of this was anyone's fault and we needed to change our mindset immediately about Julia no matter what. We needed to acknowledge that Julia is ours, and we know we love her, and will always love her and do our best for her no matter what. There was only one direction to take, and that was forward. That's just the way it was going to be. We agreed that was the case.

Ultimately, after Julia had several CTs and MRIs, and we connected with an amazing medical team at Gillette and the University of Minnesota, it was determined that she has polymicrogyria (a condition where there are too many folds in the brain, and they are unusually small). The cause was an infection from a common virus (CMV) when Julia was in utero, one that Erin had no immunity against. This infection happened at a critical time in Julia's development and changed the way her brain developed. All of that is the root cause of Julia's cerebral palsy.

While that information helped us understand Julia's overall case, it didn't change the fact that we needed to create a plan to deal with Julia the person, our beautiful daughter, and how she experiences the world. We already knew we needed to deal with the symptoms. We already knew she'd need regular speech therapy, physical therapy, occupational therapy, PM&R appointments—and whatever else to keep the doors open for her development. We already knew our lives were forever changed and we'd need to adapt to be the best advocates we could be for Julia.

Julia's diagnosis would prove to be very important for how Erin navigated her four-and-a-half-year journey with brain cancer (much longer than the typical glioblastoma prognosis of 12 months). Being there for Julia was such a motivating factor for Erin. At top of mind—beyond her own cancer treatments, radiation appointments, surgeries, and recoveries—were our girls, especially the things we needed to do for them. For Julia, she would regularly look for used medical equipment,

stay on top of therapy appointment scheduling, work on feeding, think of questions we needed to ask, think about Julia's future and what she needed—and just physically give her presence, time, and attention. Julia was not a distraction from Erin's diagnosis; she was a mission, a reason to live. Julia is nonverbal, but she would respond to her mom's love in such expressive ways. With her smile. With her laugh. With her body language. With her whole being.

What our family learned through it all is that we have a lot to learn. We need to remain open and honest. We need to listen to Julia in the form of how she responds to treatments and is developing. We need to be motivated by her attitude, her tenacity, and her infectious smile. We also learned that while there is so much *to* do, there is only so much a person *can* do. Julia's diagnosis and Erin's diagnosis taught us to look forward. To do the things we can. To control the things we can control and, hopefully, realize our own boundaries and the things that are out of our control. We learned we need to be fierce and fair advocates for Julia and kids like her—making sure they are not just surviving but are being given opportunities to thrive on their terms.

Today, Julia is loving kindergarten in her wheelchair, in her gait trainer, with her speech device, and with her friends. She is so happy to be there. It's a milestone that Erin desperately wanted to see and experience here on Earth. It's something I know Julia's infectious smile and laugh—making all those other kids smile and laugh—would warm her heart. It warms my heart, and I can't wait to see what's next for Julia.

John, father of 17-year-old David, from Luxembourg

One day when we were waiting at the airport for the wheelchair assistance to board our plane, an elderly lady looked at our son, David, who has cerebral palsy and was about 12 at the time, then turned to us and asked if we had any other children. "No," we answered. "I'm so sorry for you," she said. Her embarrassed husband told her to show more consideration. But the old lady only said what many people think when they see the family of a child with spastic quadriplegia.

Fifteen percent of the world's population has some form of disability, but few have to deal with a challenge as complex and all-consuming as spastic quadriplegia. After an intensive start, parenting for most people gets easier as their children grow in ability and autonomy. But for those who have a child with spastic quadriplegia, that period of intensive parenting never ends. A person of any age who cannot eat, drink, dress, shower, use the toilet, or perform other basic functions independently requires the same level of assistance as an infant, even if in terms of intellect and spirit they are far more capable. They can also be more demanding.

Our own story began as most cases of cerebral palsy do: with a period in the neonatal intensive care unit. We do not know what caused the problem, but we do know that the placenta had calcified and our son David had to be brought into the world by cesarean section a month earlier than planned. We have lingering doubts about the way he was handled in the hours following the birth and whether this caused or exacerbated the brain injury, but it's too late to determine this now.

When David was about six months old, his pediatrician voiced concern about our son's development and sent us to a neurologist, who said that he was just tense. We visited a physiotherapist who specialized in working with children and who told us that, on the contrary, there was something not at all right about his development. At the time, it was the middle of the summer and difficult to get a doctor's appointment. So we started doing our own research and came to the conclusion that David has cerebral palsy. This initial experience taught us to be confident about educating ourselves in our son's condition and to value the opinions of all the different professionals working with our son.

Luxembourg is a small country with a population of a bit more than half a million when our son was born. On average, only two babies with spastic quadriplegia were born there each year, so it's not surprising that there was no network of parents for us to turn to (we have since remedied that). We started to look to other countries for information (I have Irish and British nationality, and my wife is Portuguese). That is how I became involved in the International Cerebral Palsy Society.

We started visiting the specialized cerebral palsy center in Lisbon. Compared with Luxembourg, one of the richest countries in the world

with very generous social security, Portugal is a poor cousin. The public health system is underfunded and people often have to fundraise to buy mobility equipment. Yet the concentration of professionals from different disciplines working together in the CP center, together with the number of children they treat, gives them a level of expertise far higher than we found in Luxembourg. Money can certainly help, but it isn't the only answer to getting the best treatment and advice for your child with spastic quadriplegia.

The old lady at the airport did not know David. If she had, instead of seeing a helpless boy in a wheelchair, she would have recognized a determined, ambitious, positive-minded youngster who will not let any obstacle get in his way. David does not feel sorry for himself because he uses a wheelchair (he is GMFCS level IV). That is just his baseline in life, and from there he sees every new piece of mobility equipment, assistive technology device, or inclusive activity as another opportunity to live life to the fullest.

In *A Midsummer Night's Dream*, Shakespeare tells the story of a "changeling boy." According to folklore, children with disabilities—changelings—are switched by fairies for able-bodied children. Yet rather than suffer abuse, which was the sad fate of many changelings, the boy in Shakespeare's play is desired and lavished with love. That's been our experience with our David.

Parents of a child with any disability, especially spastic quadriplegia, will go through a grieving process for the child they will not have. Looking after a child with a disability is hard work, and many parents find it difficult to cope. But with all the challenges, we have also gained fortitude, resilience, a different perspective on the world, and a deeper understanding of parental love. That is a gift.

Index

Abbreviations used in index: CP cerebral palsy; ICF International Classification of Functioning, Disability and Health; ROM range of motion. Figures and tables indicated by page numbers in italics.

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Spastic quadriplegia is a 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 quadriplegia is also known as bilateral spastic CP or, simply, bilateral CP.

Spastic quadriplegia affects all four limbs and the trunk. It is generally a severe form of CP that impacts many aspects of a person's life. This practical guide addresses both the motor and nonmotor aspects of spastic quadriplegia across the lifespan and their treatment. It also includes the lived experience of families.

The writing of *Spastic Quadriplegia* was led by Marcie Ward, MD, Pediatric Rehabilitation Medicine Physician at Gillette Children's, a world-renowned center of excellence for the treatment of brain, bone, and movement conditions. *Spastic Quadriplegia* is part of 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 Quadriplegia*.

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