"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

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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.

* 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.

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.

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.

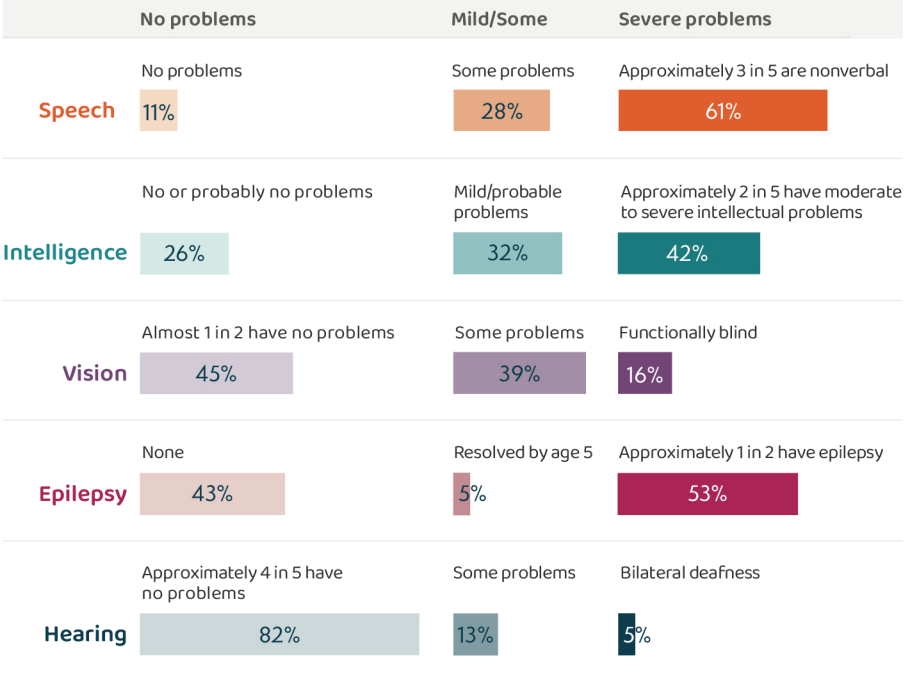

Figure 2.1.2 Prevalence of associated problems among children age five with spastic quadriplegia (all GMFCS levels). Data also includes children with triplegia.

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.

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 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.

* 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.

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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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