Appendices: Spastic Quadriplegia

Appendix 1

Measurement tools

Table A1.1 lists examples of some commonly used measurement tools in CP.

Table A1.1 Measurement tools used in CP

Measurement tool	**Variable**
DAYC (Developmental Assessment of Young Children)	Cognition, communication, social-emotional development, physical development, and adaptive behavior
Peabody Developmental Motor Scales	Gross and fine motor function
Bayley Scales of Infant and Toddler Development	Cognition, communication, social-emotional development, physical development, and adaptive behavior
Goniometer	ROM of joint
GMFM (GMFM-88 and GMFM-66)	Gross motor function
10-meter or 6-minute walking test	Walking
Gillette Functional Assessment Questionnaire (FAQ)	Functional mobility
Functional Mobility Scale (FMS)	Functional mobility
Gait Outcomes Assessment List (GOAL)	Gait priorities and functional mobility
Pediatric Outcomes Data Collection Instrument (PODCI)	Pediatric health
PEDI-CAT	Daily activities, mobility and social/cognitive function
Canadian Occupational Performance Measure (COPM)	Self-perception of performance in everyday living
CP CHILD	Health, comfort and well-being and caring for the child's needs

Appendix 2

Bones, joints, muscles, and movements

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

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

- A lever (a rigid bar)
- A fulcrum (a point about which the lever pivots)
- A resisting force (or load, such as a weight to be moved)
- An applied force (or effort; something that is doing the moving) (see Figure A2.1)

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

- The forearm bones are the lever.
- The elbow joint is the fulcrum.
- The object being lifted is the resisting force.
- The contraction of the elbow flexor muscles creates the applied force.

Muscles provide the action; the bones just follow.

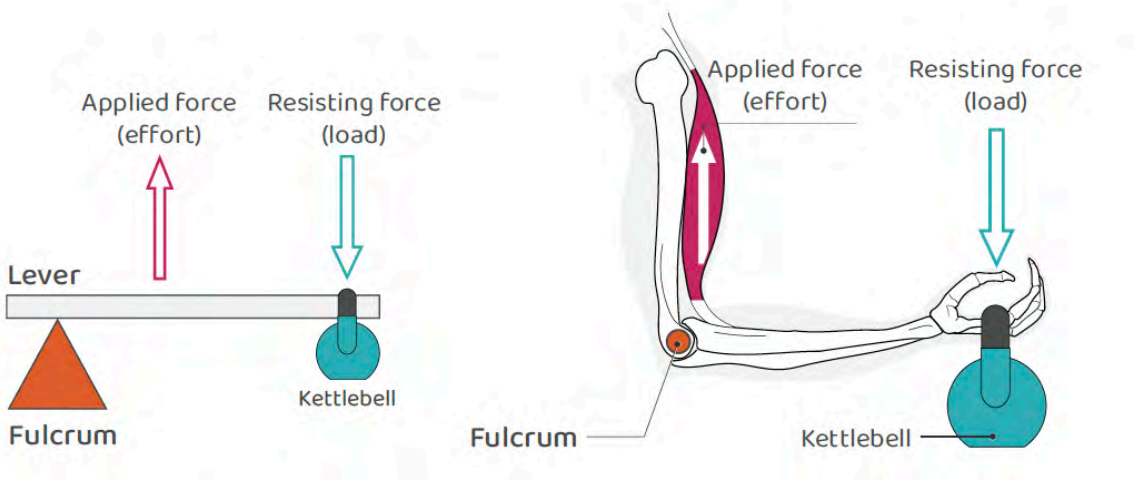

Figure A2.1 A lever (left) and the corresponding parts in the human arm (right).

Both the resisting force and applied force act on the lever at a distance from the fulcrum, which creates a torque or rotation (also called a "moment") about the fulcrum. This distance is called the force's "lever arm" (or "moment arm"). Even if the force stays constant, when the lever arm increases in length, the torque increases, and vice versa. Using the example in Figure A2.1, if the kettlebell were moved closer to the fulcrum (elbow), effectively shortening the lever arm, a smaller applied force (i.e., muscle contraction) would be required to lift it.

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

There are three types of muscles in the body:

- Cardiac: Muscle that forms the bulk of the wall of the heart

- Smooth: Muscle located in the walls of hollow internal structures such as the blood vessels, stomach, and intestines
- Skeletal: Muscle attached (mostly) to bones

Spastic quadriplegia primarily affects skeletal muscle.*

Skeletal muscles contract to produce movement or maintain posture. The bones cannot stand up on their own; gravity would pull them down. When muscles contract, in addition to causing movement, they exert force, which keeps the body erect. Without these forces opposing gravity, the bones would collapse in a heap on the ground. In a sense, the bones are like the limbs of a marionette (or puppet, see Figure A2.2). The marionette cannot stand up on its own.

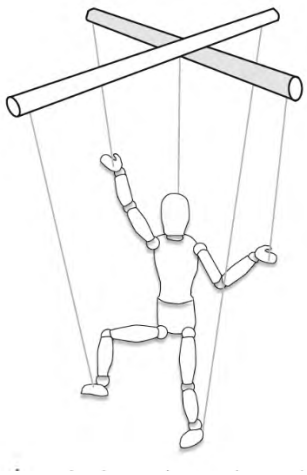

Figure A2.2 A marionette (puppet).

There are three types of muscle contractions:

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

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

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

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

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* There are some reports of smooth muscle being affected in CP.[1,2,3]

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

Something else to consider is range of motion (ROM), also called "range of movement," which is a measure of joint flexibility. The actual ROM through which a joint can be passively moved is measured in degrees. An instrument called a goniometer* is used to measure the ROM of a joint. (See Figure A2.3). A video about measuring ROM is included in **Useful web resources.**

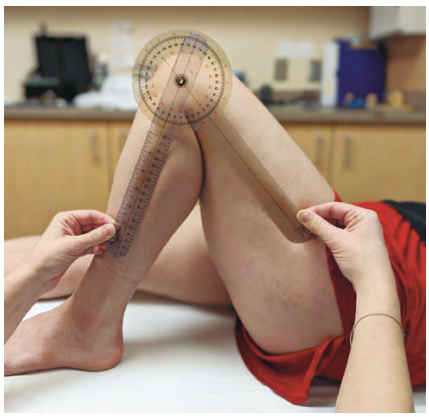

Figure A2.3 Measuring the ROM of the knee joint using a goniometer.

Tables A2.1 and A2.2 explain the movements, joint ROMs, and key muscles for both the upper and lower limbs. These tables are included as a reference and may be helpful at different times; for example, it may be useful to take them to some appointments. Below are some relevant points:

- Muscles are generally arranged in pairs around a joint. The muscles on one side of the joint move the joint in one direction, while the muscles on the other side of the joint move the joint in the opposite direction. Key muscles are identified at each joint, but minor muscles have not been included.
- Movements typically affected by spasticity are shown on the left side of the tables and are indicated with a green background. Some, but not necessarily all, of the muscles responsible for those movements may be affected by spasticity. The tables show the movements typically affected by spasticity, but there may be some variation between individuals.
- Two-joint muscles play a role in movement at two joints. (Some muscles in the hand and foot cross more than two joints.) The most significant movements affected by two-joint muscles and spasticity are indicated in orange on the left side of the tables.
- Typical ROMs for each joint are shown. The closer a joint's ROM is to typical, the better. A muscle contracture is a limitation of a joint's ROM.[4] The terms "muscle contracture" and "tight muscle" are used interchangeably in the CP field and in this book.

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

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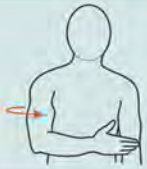
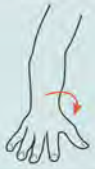
* A goniometer is like a movable protractor, used for measuring angles, as shown in Figure A2.3.

Table A2.1 Upper limb movements, joint ROMs,[5] and key muscles

MOVEMENT (Green background indicates movements affected by spasticity *on the involved side* in hemiplegia)		KEY MUSCLES RESPONSIBLE FOR THE MOVEMENT (Two-jointed muscles are indicated in orange)
Shoulder adduction Movement of the arm toward the middle of the body (midline) **ROM 0 to 140 degrees**		**Shoulder adductors** • Latissimus dorsi • Teres major • Pectoralis major
Shoulder flexion Movement of the arm upward toward the face **ROM 0 to 180 degrees**		**Shoulder flexors** • Pectoralis major • Deltoid
Shoulder internal rotation Movement of the upper arm internally toward the middle of the body (midline) **ROM 0 to 70 degrees**		**Shoulder internal rotators** • Latissimus dorsi • Teres major • Pectoralis major
Elbow flexion Movement of the forearm toward the upper arm **ROM 0 to 150 degrees**		**Elbow flexors** • Biceps • Brachialis • Brachioradialis
Forearm pronation Internal rotation of the forearm that results in the hand moving from the palm-up to the palm-down position **ROM 0 to 80 degrees**		**Pronators** • Pronator teres • Pronator quadratus
Wrist flexion Movement of the palm of the hand toward the inside of the forearm **ROM 0 to 80 degrees**		**Wrist flexors** • Flexor carpi radialis • Flexor carpi ulnaris • Palmaris longus

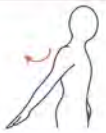
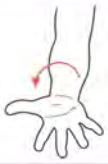
OPPOSITE MOVEMENT		KEY MUSCLES RESPONSIBLE FOR THE OPPOSITE MOVEMENT
Shoulder abduction Movement of the arm away from the middle of the body (midline) **ROM 0 to 45 degrees**		**Shoulder abductors** • Supraspinatus • Deltoid
Shoulder extension Movement of the arm to the back of the body **ROM 0 to 60 degrees**		**Shoulder extensors** • Deltoid • Latissimus dorsi • Teres major
Shoulder external rotation Movement of the upper arm externally away from the middle of the body (midline) **ROM 0 to 90 degrees**		**Shoulder external rotators** • Infraspinatus • Teres minor
Elbow extension Movement of the forearm away from the upper arm **ROM 0 to 150 degrees**		**Elbow extensors** • Triceps
Forearm supination External rotation of the forearm that results in the hand moving from the palm-down to the palm-up position **ROM 0 to 80 degrees**		**Supinators** • Supinator
Wrist extension Movement of the palm of the hand away from the inside of the forearm **ROM 0 to 70 degrees**		**Wrist extensors** • Extensor carpi radialis • Extensor carpi ulnaris • Extensor carpi radialis brevis

Table A2.1 Continued

MOVEMENT (Green background indicates movements affected by spasticity *on the involved side* in hemiplegia)	KEY MUSCLES RESPONSIBLE FOR THE MOVEMENT (Two-jointed muscles are indicated in orange)	OPPOSITE MOVEMENT	KEY MUSCLES RESPONSIBLE FOR THE OPPOSITE MOVEMENT
Thumb adduction Movement of the thumb toward the fingers **ROM 0 to 80 degrees**	**Thumb adductors** • Adductor pollicis	**Thumb (radial) abduction** Movement of the thumb away from the fingers **ROM 0 to 80 degrees**	**Thumb abductors** • Abductor pollicis longus • Abductor pollicis brevis
Thumb flexion Movement of the thumb into palm **ROM 0 to 50 degrees**	**Thumb flexors** • Flexor pollicis longus • Flexor pollicis brevis	**Thumb extension** Movement of the thumb away from the palm **ROM 0 to 50 degrees**	**Thumb extensors** • Extensor pollicis longus • Extensor pollicis brevis
Finger flexion Movement of the fingers toward the palm **ROM 0 to 90 degrees**	**Finger flexors** • Flexor digitorum superficialis • Flexor digitorum profundus	**Finger extension** Movement of the fingers away from the palm **ROM 0 to 90 degrees**	**Finger extensors** *Individual muscles not listed*

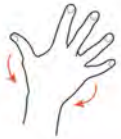


Table A2.2 Lower limb movements, joint ROMs, [6,7] and key muscles

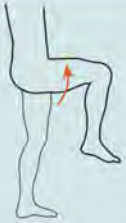
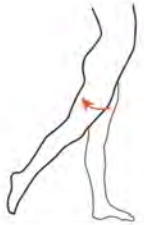
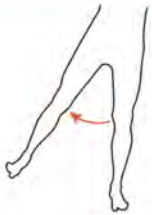
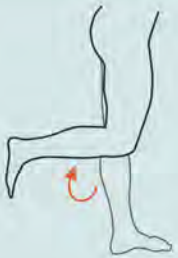
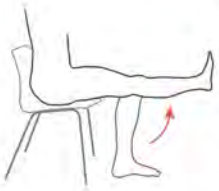
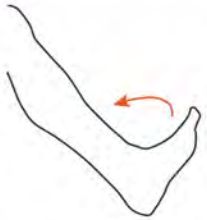
MOVEMENT (Green background indicates movements affected by spasticity)		KEY MUSCLES RESPONSIBLE FOR THE MOVEMENT (Two-jointed muscles are indicated in orange)	OPPOSITE MOVEMENT		KEY MUSCLES RESPONSIBLE FOR THE OPPOSITE MOVEMENT
Hip flexion Movement of the thigh up toward the pelvis **ROM 0 to 125 degrees**		**Hip flexors** • Iliopsoas • Rectus femoris	**Hip extension** Movement of the thigh away from the pelvis **ROM 0 to 10 degrees**		**Hip extensors** • Gluteus maximus • Hamstrings
Hip adduction Movement of the thigh toward the midline **ROM 0 to 20 degrees**		**Hip adductors** • Adductor longus • Adductor magnus • Adductor brevis • Gracilis	**Hip abduction** Movement of the thigh away from the midline **ROM 0 to 45 degrees**		**Hip abductors** • Gluteus medius
Hip internal rotation Rotary movement of the thigh toward the midline; also known as inward or medial rotation **ROM 0 to 45 degrees**		*Individual muscles not listed*	**Hip external rotation** Rotary movement of the thigh away from the midline; also known as outward or lateral rotation **ROM 0 to 45 degrees**		*Individual muscles not listed*
Knee flexion Increasing the angle between the thigh and lower leg **ROM 0 to 140 degrees** Note: Reference point is the straight leg. The angle increases the nearer the lower leg moves to the thigh.		**Knee flexors** • Hamstrings • Gastrocnemius	**Knee extension** Decreasing the angle between the thigh and lower leg **ROM 140 to 0 degrees** Note: Reference point is the flexed leg. The angle decreases the further the lower leg moves away from the thigh.		**Knee extensors** The quadriceps (quads) consist of four muscles: • Rectus femoris • Vastus intermedius • Vastus lateralis • Vastus medialis

Table A2.2 Continued

MOVEMENT (Green background indicates movements affected by spasticity)	KEY MUSCLES RESPONSIBLE FOR THE MOVEMENT (Two-jointed muscles are indicated in orange)	OPPOSITE MOVEMENT	KEY MUSCLES RESPONSIBLE FOR THE OPPOSITE MOVEMENT
Ankle plantar flexion Movement of the foot away from the lower leg **ROM 0 to 45 degrees** Note: Reference point is the 90-degree angle between the lower leg and the foot. 	**Ankle plantar flexors** • Gastrocnemius • Soleus	**Ankle dorsiflexion** Movement of the foot toward the lower leg **ROM 0 to 20 degrees** Note: Reference point is the 90-degree angle between the lower leg and the foot. 	**Ankle dorsiflexors** • Tibialis anterior • Toe extensors

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Appendix 3

Epilepsy management

Epilepsy management is complex. Epilepsy may evolve over time as the individual gets older, so the evaluation of the condition and its management is ongoing. Since clinical expertise can vary, it is important to know that information about management in this book may be different to practice at different hospitals and treatment centers. Management is not "one size fits all"; it must be customized.

The main goal of epilepsy management is to *prevent, reduce,* or *stop* seizures. Some related important terms to understand include:

- **Seizure control:** Effective epilepsy management that results in a decrease in frequency, severity, and/or duration of seizures.
- **Seizure freedom:** A set period without any seizures; the ultimate goal of epilepsy management.
- **Remission:** A state where an individual with epilepsy is seizure-free for at least six months.
- **Resolved:** A state where an individual with epilepsy has remained seizure-free for the last 10 years, with no antiseizure medications for the last 5 years, or the individual had an age-dependent epilepsy syndrome and is past the applicable age for this diagnosis (i.e., self-limited neonatal or infantile epilepsy syndromes).

Why manage epilepsy?

Management of epilepsy is important for the following reasons:

- **To protect the brain from damage:** Epileptic seizures may lead to damage of areas in the brain, especially when they are prolonged or uncontrolled.
- **To protect organs and body systems from damage:** Epileptic seizures (especially those with motor signs) may lead to injuries and lesions in various body organs (e.g., kidneys or liver), or body systems (e.g., cardiovascular or musculoskeletal systems).
- **To prevent status epilepticus:** This condition, in which seizures last more than five minutes or occur in close succession (one after the other, without a return to baseline), is life-threatening.
- **To prevent SUDEP (sudden unexpected death in epilepsy):** This rare complication of epilepsy is named to describe the death of an individual with epilepsy when no other cause of death can be found.
- **To ensure safety and prevent injury:** Individuals with epilepsy are at an increased risk of accidental injuries from falls, motor vehicle accidents, and accidents around water, fire, and in other activities.
- **To improve quality of life:** Seizure control correlates with the ability to participate fully in life, including social activities, physical activities, education, employment, driving, and independent living.

How is epilepsy managed?

The management of epilepsy generally includes:

- **Pharmaceutical treatments**, involving the use of antiseizure medications, either as monotherapy (one medication) or polytherapy (more than one medication)
- **Non-pharmaceutical treatments**, involving the ketogenic diet, neuromodulation (repetitive electrical discharges administered through a device), and epilepsy surgery
- **Other medications or supplements**, including vitamins or medical cannabis

Pharmaceutical treatments are generally tried first. However, some epilepsy syndromes and drug-resistant epilepsy are best managed with non-pharmaceutical treatments or other medications or supplements. Pharmaceutical treatments, non-pharmaceutical treatments, and other medications or supplements can be used with the same individual and at the same time.

Management options for epilepsy are shown in Table A3.1.

Table A3.1 Management options for epilepsy

Management	Description	Indications for use
Pharmaceutical treatments		
Monotherapy	One antiseizure medication (may try a different medication if the first doesn't work)	All types of epilepsy, generally tried first
Polytherapy	More than one antiseizure medication given at the same time (may try different combinations)	All types of epilepsy, when monotherapy does not work
Non-pharmaceutical treatments		
Ketogenic diet	Specialized diet with a very low amount of carbohydrates	Used when polytherapy does not work, or when the epilepsy type, epilepsy cause, or epilepsy syndrome is more responsive to non-pharmaceutical management
Neuromodulation	Repetitive electrical discharges administered through a device (for the management of epilepsy, these devices are surgically implanted)	
Epilepsy surgery	Surgery to areas of the brain where seizures are thought to start or spread to	

Table A3.1 Continued

Management	Description	Indications for use
Other medications and supplements		
Medications	Medications other than antiseizure medication include: • Immunotherapies (treatments that alter the immune system) • Steroids (medications with anti-inflammatory properties) • ACTH (a type of hormone therapy).	Used when the epilepsy type, epilepsy cause, or epilepsy syndrome is known to be responsive to a particular medication
Vitamins	Dietary supplements	Used in epilepsy syndromes known to be responsive to a particular vitamin
Medical cannabis	A pharmaceutical form of the cannabis plant	Used in epilepsy types and epilepsy syndromes known to be responsive to cannabis

More information on epilepsy is available in the book *Epilepsy* in the **Gillette Children's Healthcare Series.**

Appendix 4

Scoliosis management

Treatment options for scoliosis can range from nonsurgical methods, such as observation with repeat X-rays, to surgical methods, such as spinal fusion. Treatment options include the following (note that goals are shown in italics):

- Observation: Regular spine X-rays and clinical exams with a spine specialist *to monitor scoliosis curve for possible progression.*
- Bracing: A spinal brace that applies corrective forces to the spine *to slow or stop scoliosis curve progression.*
- Surgery: Surgery performed *to prevent future progression and improve the scoliosis curve (decrease the Cobb angle).* There are many types of scoliosis surgery. The most common type is spinal fusion, defined as fusing (joining together) two or more vertebrae in the spine; screws and metal rods are typically used to hold the spine in the straightened position and facilitate fusion between bones.

Table A4.1 summarizes these treatment options

Table A4.1. Treatment options for managing scoliosis

Treatment options	Indications	Goals
Observation		
Skeletally immature	Cobb angle less than 20 degrees	Monitor scoliosis curve through repeat X-ray images for possible progression
Skeletally mature	Cobb angle between 30 and 50 degrees	
Bracing		
Skeletally immature	Cobb angle between 20 and 45 degrees	Slow or stop scoliosis curve progression Prevent or delay surgery
Skeletally mature	Not an appropriate treatment once skeletally mature	
Surgery		
Skeletally immature	Cobb angle greater than or equal to 50 degrees	Stop curve progression Improve the spinal curve (decrease the Cobb angle)
Skeletally mature	Cobb angle greater than 50 degrees	

		Achieve a balanced spine and posture

More information on scoliosis management in CP is available in the book *Congenital, Neuromuscular, Syndromic, and Other Causes* in the **Gillette Children's Healthcare Series**.