CLIENTS WITH LOW MUSCLE

Etiology, Issues and Wheelchair Seating Challenges

INTRODUCTION

Low muscle tone—most of us recognize it when we see or feel it, but where is it coming from and what is really going on? Physicians and therapists agree on the difficulty of assessing low tone. It can be difficult to diagnose and determine the source of the problem, especially in young children. A deeper understanding of low tone symptoms, causes, possible complications and postural tendencies will advance our ability to provide optimal seating for clients who present with this issue.

DEFINITION

Hypotonia is the medical term that describes low or decreased muscle tone. Low tone is a symptom of an underlying condition, not a diagnosis by itself. People with hypotonia have a central nervous system injury or condition, a genetic or chromosome disorder, or a problem in the muscles themselves. These conditions can be divided into four broad categories—central nervous system, peripheral nerve, neuromuscular junction and muscle disorders.

Hypotonia is usually diagnosed in children but also develops in adults usually after an illness, injury or as part of a disease process. When no clear etiology is determined in children it is called idiopathic or benign congenital hypotonia. But the term "benign" is controversial in some circles because what appears nonthreatening at an early age may later develop into a serious condition. Understanding the cause of low tone, with the prognosis for progression or improvement, has a large impact on management of the condition, including seating and wheeled mobility decisions.

Muscle tone is defined as the natural tension in a resting muscle—its ability to contract in response to external stretch. Skeletal muscle tone is not muscle strength. Muscle strength is the force generated by a muscle group in a single contraction against resistance, often measured with manual muscle testing or devices like dynamometers. Weaker muscle groups generate reduced force when they contract. Muscular endurance must also be considered and is different than pure strength. Endurance is the ability of a muscle group to exert less than maximal force for extended periods of time—to keep doing something over and over. A good example of this in a functional context is manual wheelchair propulsion. A person with sufficient motor control and strength to push for a short distance needs adequate muscular endurance to carry on for the long haul and functionally propel a

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TONE

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manual wheelchair. A person with high or low tone can have varying degrees of weakness and muscular endurance. Depending on the condition, developing increased strength and endurance, regardless of tone, is a possibility.

A healthy muscle is never fully relaxed but always retains some tension or mild contraction even when not being used in a specific activity. For instance, normal tone in our trunk and neck muscles stabilizes our upright sitting posture. Without it we would collapse, as evidenced by what happens when one falls asleep sitting up. Muscle tone relaxes as a normal part of sleep, so if we fall asleep while sitting up we will often wake with our head flopped forward or to the side—and likely a stiff neck!

The term "low tone" is frequently used interchangeably with muscle weakness, but it is important to differentiate between the two. While hypotonia is often associated with muscle weakness, they are not the same thing. A person with low resting tone may compensate and improve functionally over time by strengthening muscle groups. This concept is relevant when assessing clients with hypotonia for seating and wheeled mobility. Over time, an effective seating intervention may assist a person with low tone in strengthening and building endurance in particular muscle groups that will improve function—for instance head control. It does not change the underlying muscle tone, but it can decrease its functional impact.

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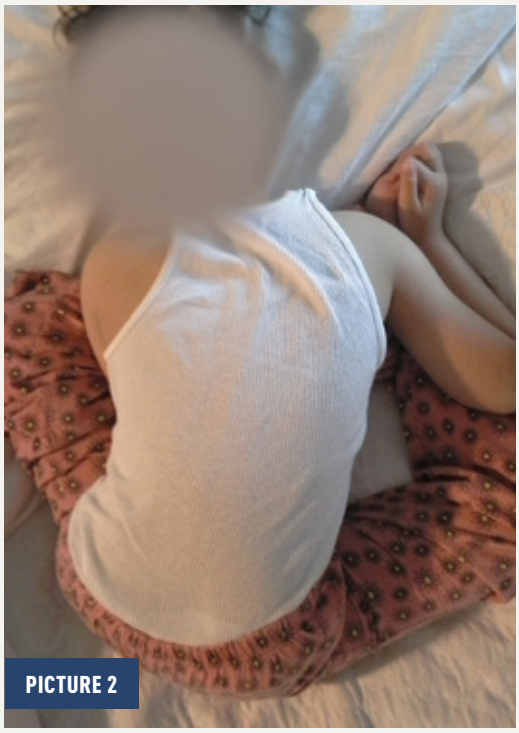
PICTURE 1

Picture 1: An extreme example of sliding out with associated posture problems when the pelvis is not stabile.



PICTURE 2

Picture 2: Excessive flexibility allows this kind of sleeping position.

Understanding the differences between normal, high and low tone is essential because a person in each category will respond differently to the same seating intervention. Normal tone allows a range of movement that supports stability and motor control while allowing both activity and rest. Muscles with normal tone feel firm but yielding. There is a range of strength amongst individuals, but we are talking about the resting tension of the muscle—its potential to contract.

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Hypertonia (high tone) describes excessive resistance to passive stretch. In a resting state, range is lessened, and the muscles feel tight or rigid, they have reduced capacity to stretch. High tone can occur with or without spasticity. Spasticity is velocity dependent resistance to passive movement—a quick stretch elicits increased resistance. The Ashworth Scale and deep tendon reflex testing are often used to assess spasticity.

Hypotonic muscles feel soft and sometimes even mushy. There is reduced or even no resistance to passive movement. The muscle contracts very slowly and may be unable to sustain contraction for long, therefore always remaining somewhat stretched out depending upon the severity of hypotonia. Lax ligaments are common with low tone and result in excessive joint flexibility or hypermobility. These people lack core stability and may feel like they will slip through your hands during manual transfers between support surfaces. In fact, the "shoulder slip through" test is one component of assessing young children with low tone, carried out by holding the trunk under the arms and suspending a child vertically. Interestingly, a person may have mixed presentation of hypotonia and spasticity. This can be seen in people born with low tone, for instance secondary to a chromosome disorder underlying another condition. This author has seen individuals with Down syndrome and traumatic brain injury, for instance, or cerebellar hypotonia and hemiparesis with some spasticity.

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Hypotonia is sometimes a transient finding, as in babies who are born too early and have not developed normal tone in utero. Low tone can emerge as part of a specific disease process or injury. Some people have mixed tone: for instance, individuals with cerebral palsy may have high tone in some parts of their bodies with low underlying trunk tone. A person may have very low tone on one side and normal tone on the other after a cerebral vascular accident (CVA). In others, tone changes over time, becoming higher or lower as conditions evolve or resolve.

There is no universally accepted scale for measuring hypotonia. Much of the literature focuses on evaluation of hypotonia in young children, with exploration of non-invasive, clinical assessments. Hypotonia can be caused by more than 500 genetic disorders. Determining the cause is important for management of the condition and with respect to genetic testing for inherited syndromes. A combination of personal and family history, as well as developmental and clinical testing with recognition of characteristics, are typically used in assessing low tone. Procedures used by therapists and pediatricians include testing of suspended prone anti-gravity responses, the "shoulder slip through" test, and neurologic assessment of tone, reflexes and strength. By nature, these tend to be subjective, but Govender (2016) has proposed a prototype algorithm for clinical assessment of children with hypotonia to provide more objective decision-making.

CENTRAL VS. PERIPHERAL HYPOTONIA

To function normally, muscles depend on information from motor nerves. Motor commands to the muscles are generated in the motor region of the cerebral cortex or the brainstem in the central nervous system. The upper motor neurons do not leave the central nervous system (brain and spinal cord) but synapse with lower motor neurons that conduct messages to the muscles in the rest of the body. Origins of hypotonia are first differentiated as being central or peripheral.

Central hypotonia is most common, accounting for about 66 to 88 percent of cases in children (Harris, 2008). Central hypotonia involves upper motor neurons, meaning that nerve signals are disrupted in the brain or spinal cord. The messages given to the lower motor neurons are atypical and those messages are then passed on to the muscles, resulting in decreased muscle tone. Typical causes are damage or developmental problems in the brain or spinal cord such as malformations, lack of oxygen or bleeding in the brain, serious infections like meningitis or encephalitis that cause brain damage, spinal cord injury, and genetic or metabolic disorders affecting the central nervous system (Bay & Johnston, 2016). Weakness is often, but not always, associated with central hypotonia—thus the possibility of strengthening to compensate for low tone sometimes exists. In central hypotonia, seizures, developmental delays, dysmorphic facial features, micro or macrocephaly, abnormal eye movements, and other signs of central nervous system dysfunction are often seen. Diagnoses manifesting central hypotonia include cerebral palsy (CP) (hypotonic CP being much less common than other forms), CVA, chromosome disorders (i.e. Prader-Willi, Down, Phelan-McDermid syndromes), metabolic diseases (i.e. disorders of glycosylation,

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AADC deficiency, or underdeveloped endocrine glands), spinal cord injuries and multiple sclerosis.

Peripheral hypotonia is less commonly seen in children but is a more common cause of later developing low tone in adults. It involves lower motor neurons, with interference occurring between the spinal cord and the muscles. The messages sent from the brain may be accurate and intact but cannot be transmitted properly to the muscles for some reason. Peripheral hypotonia is most often associated with significant muscle weakness. There are three issues typically related to this type of low muscle tone: (1) damage between the spinal cord and muscle, (2) poor communication between the end of the nerve and the muscle, and (3) problems in the muscle itself. Some examples are: (1) disorders in which the myelin coating of nerve fibers is damaged, interfering with transmission of information to the muscles (i.e. Charcot-Marie-Tooth disease), (2) disruption at the neuromuscular junction, where the nerve directly communicates with the muscle (i.e. myasthenia gravis), (3) muscle diseases that interfere with essential processes for forming and maintaining healthy muscles (i.e. muscular dystrophies), (4) motor neuron diseases in which the nerves are progressively damaged, causing muscle wasting (i.e. spinal muscular atrophy), and (5) disorders of connective tissue that binds and supports other tissues (i.e. Ehlers-Danlos syndrome).

Some conditions exhibit combined central and peripheral hypotonia; examples are giant axonal neuropathy (GAN) and amyotrophic lateral sclerosis (Lou Gehrig's disease).

Diagnostic tools for determining the origin of hypotonia include blood tests, EMGs, EEGs, spinal taps, CT scans, MRIs, and muscle biopsies. In young children, differences in head lag when pulled to sit, supine lying posture (hips widely abducted), deep tendon reflexes, prone anti-gravity responses, and cognition/affect are clinically valuable in determining the origin when other diagnostic tools are not available or inconclusive.

WHAT DOES HYPOTONIA LOOK LIKE?

People with hypotonia show certain typical characteristics, varying according to the underlying condition. Low tone can be mild or very severe. People with mild hypotonia can typically ambulate. They have developed coping mechanisms over time that help them function, but the quality of their movement and posture is atypical. They often sacral sit with rounded back, have overly flexible joints, and may have poor balance with tendency toward clumsiness and frequent falling. Compensatory strategies often help with difficulty getting up from sitting or lying, and may not be noticeable to an untrained eye. Thus, these individuals may go through life unaware they have low tone until it comes to the forefront. For instance, mothers with myotonic dystrophy can be functional and unaware of their condition until they give birth to a child who is much more seriously affected.

THOSE WHO REQUIRE SEATING AND WHEELED MOBILITY SERVICES WILL SHOW MANY OF THE FOLLOWING PHYSICAL CHARACTERISTICS IN VARYING COMBINATIONS AND DEGREES:

- Poor pelvic and trunk stability/control—this compromises the foundation of stable sitting posture. People with hypotonia often sit with a posterior pelvis, rounded back and trunk collapsed forward to gain stability. But without a secure pelvis, they will slide out of the chair (see Picutre 1). If the pelvis but not the upper body is stabilized, the person will often fall forward with an anterior pelvic tilt, compensating with lumbar lordosis to remain upright and avoid collapsing. Another scenario is an oblique or rotated pelvis to gain stability, but this contributes to an unstable trunk and compensatory spinal curve leading to or worsening scoliosis. Eventually this can progress to rib cage/pelvis proximity with associated breathing and other health complications.

- Poor head control—compromised pelvic and trunk stability leads to excessive neck flexion or hyperextension; this compromises the visual field, swallowing, and breathing. In some cases, a lateral head posture becomes prominent. In all these examples, overstretching with shortening of soft tissues on the opposing side can eventually limit passive neck range and cause chronic pain.

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• Poor weight bearing through legs and/or shoulders—weight bearing through the legs is difficult or impossible as is using the upper limbs to support trunk posture. It is crucial to support these body parts to prevent injuries like shoulder subluxation and foot drop secondary to habitually dependent positions.

• Diminished reach and grasp—reduced active movement and grip strength affects upper extremity use for activities including wheelchair mobility.

• Shallow breathing—respiration is often affected, especially in peripheral hypotonia, with up to 50 percent of people ages 3-55 requiring some form of breathing assistance at night and others requiring 24-hour assistance.

• Weak or quiet voice—related to limited breath support and muscle control.

• Flat affect with open mouth—when facial muscles are affected there may be drooling and lip closure problems, the person may appear "half asleep" with droopy eyelids, and expressiveness of the face overall is diminished.

• Eating/swallowing difficulties—breathing and oral motor tone/control problems include reduced tongue movement for moving food in the mouth, with chewing, lip closure and swallowing challenges. Sometimes eating orally just takes too much energy.

• Absent or slow postural reactions—maintaining stability when balance is challenged is a complex process, requiring an intact and mature nervous system. Individuals with low muscle tone often have slow or absent reactions that will impact seating decisions.

• Joint hypermobility—lax ligaments associated with low muscle tone allow more range of movement than normal (see Picture 2). This can lead to joint instability with potential for subluxation or dislocation of joints if these are not supported in a functional, well-aligned position.

This picture of physical characteristics can be used to determine specific needs for postural support during mat evaluation and how to provide it.

GRAVITY, LOW TONE AND POSTURE

Human beings have three available postural orientations—lying, sitting, and standing. Our earliest postural experiences occur in lying, progressing as we gain control against gravity to sitting and standing. Developing motor skills with gravitational challenge is a big deal and even more so for a person with low tone. We can think of it as being a "human sandwich." This describes the reality of our lives as human beings, functioning on a support surface and constantly influenced by gravity. The term was coined by Noreen Hare (http://www.hafpa.info) to describe our place in the sandwich—the filling. Whether we are lying, sitting or standing, we function in an environment where gravity plays a huge role.

During typical development, there is smooth progression through developmental stages encompassing a variety of movements; but children with low tone may be unable to counteract gravity in the first orientation of lying—much less the others. Raising head and shoulders against gravity in prone or flexing hips and reaching with arms in supine lying may be difficult or impossible. Therapeutic positioning in lying at early stages offers antigravity experiences that hypotonic children are unable to achieve on their own.

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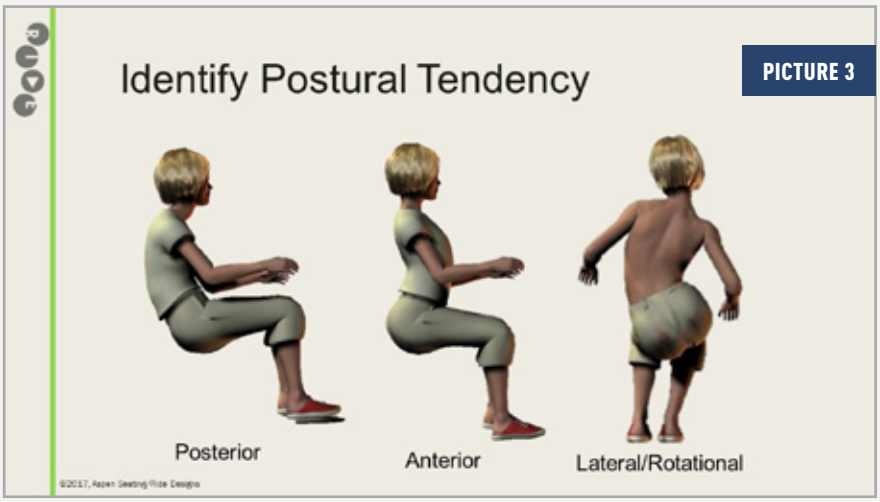
Therapeutic positioning also builds the foundation for a future of well-aligned and functional sitting, beginning with a stable lying posture. Habitual lying postures can become destructive if they are not identified and corrected early. Long hours spent lying with limbs in extreme positions secondary to lax ligaments stresses vulnerable joints. Habitually asymmetrical postures in lying can eventually lead to a distorted pelvis and spine that will complicate wheelchair seating. Positioning assistance in lying is also essential for adults with acquired hypotonia. A well-aligned, supported lying posture helps protect joints and facilitates preserving the symmetrical body with which most people are born. Attention to this aspect of positioning is often overlooked in the United States. But if it is addressed properly, it will set the stage for successful seating interventions going forward because lying and sitting are linked and will affect each other over time. In fact, the Chailey Levels of Motor Ability (Pountney et al, 2004) specifically link prerequisite lying development with achievement of beginning sitting skill in children—the ability to lie in balance being a foundation for balanced sitting.

Impaired lying balance is associated with sitting challenges. Sitting problems are more visible and extreme because the weight-bearing surface area of the body is much smaller, and the challenge of gravity is greater. Yet sitting is highly desirable for physical, social, and psychological reasons as well as mobility. In sitting, a great deal more support is necessary compared to lying to counteract the forces of gravity and natural postural tendencies.



Picture 3: Example postures associated with posterior, anterior and lateral/rotational tendencies

WHAT ARE POSTURAL TENDENCIES?

The Oxford dictionary defines tendency as "an inclination towards a particular characteristic or type of behavior." All of us have postural tendencies as humans, but the stakes are higher for wheelchair users. Here we are focusing on what typically happens in a person with low muscle tone sitting under the influence of gravity. The foundation for sitting is the pelvis, and in that context three postural tendencies occur—posterior, anterior and lateral/rotational—as illustrated in the Picture 3 example postures.

When people with low tone have a limited repertoire of posture choices, they often cannot easily independently recover their position if lost. Considering postural tendencies and their influence on seating outcomes (negative and positive) for people with low tone is essential during evaluation. Hetzel (2013) describes the importance of supporting people in a non-destructive postural tendency. Typical activities wheelchair users engage in are sitting, moving from place to place (propelling, driving or being pushed), and fine motor/functional activities like self-care and using technology. By determining the proportion of time spent doing these activities in a person's day, appropriate goals can be established during the wheelchair evaluation. For many clients sitting and doing fine motor/functional activities will win out, but in any case, understanding this balance is an essential part of the evaluation—especially for people with hypotonia for whom fatigue and endurance are often important considerations.

The posterior tendency (not to be confused with a posterior tilt/sacral sitting) is one that allows the person to rest back in a well-aligned posture without distortion of the body while sitting. The resting posture must allow the person to engage in activities, while being able to rest back in appropriate alignment for breaks. For people with low tone, fostering a posterior tendency is the non-destructive way to go, with less chance of contributing to skeletal distortions that will result in future complications, pain and dysfunction. This can be relatively simple for some clients and highly complex for others, requiring careful thought and input from all team members and especially from the wheelchair user, family and caregivers. The more information gained from the history, prognosis, mat assessment and, ideally, seating simulation during the evaluation process, the better. Our seating recommendations have long term effects, and funding challenges often preclude early equipment replacement if we don't get it right.

WHEELCHAIR FRAME STYLES AND LOW TONE

Given the powerful influence of gravity on a person with low tone, tilt-in-space and recline seat functions are important considerations in selecting a wheelchair frame. Recline was developed first and has been available for upward of 80 years on some wheeled mobility devices. Recline allows opening of the seat-to-back angle as the back moves rearward, while the seat remains in a steady orientation—like a lounge chair at the pool. This allows the person to sit with a more open hip angle, which can have good and bad consequences. Individuals with low tone may respond to an excessively closed seat-to-back angle by collapsing forward or scooting out if they are uncomfortable and strong enough. Using small amounts of recline can resolve this problem and positively influence breathing and digestion by allowing more chest and abdominal expansion. Recline can also contribute to sliding out of the chair unless specific action is taken to keep this from happening.

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Picture 4: Extreme sacral sitting and forward trunk collapse when using tilt only.

Picture 5: Using the same seating system as in Picture 4, with addition of recline, promoted more open and upright posture and reduced sliding out

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Tilt-in-space developed later, about 30 years ago in the early 1980s. Tilt-in-space is like a rocking chair—the seat and back remain at the same angle to each other, while the whole unit changes orientation relative to gravity. This means that a person who is tilted will experience weight shift rearward while their hip angle remains constant. Adjustable and fixed (non-adjustable) tilt-in-space can both influence muscle tone and provide gravitational relief to support sitting posture. Adjustable tilt is necessary for weight shifts. When using tilt, the seat-to-back angle does not open and as a result, some people have less tendency to slide out, but this is not universal. When people cannot tolerate a seat-to-back angle, they may persistently seek relief by sliding out to a more open position. This is seen in people with low as well as high tone.

In the 1990s, Medicare funded only recline wheelchairs regardless of client need. A full range of choices is available today for people with low tone, who may benefit from both tilt and recline for optimal function. Tilt allows for relief from the forces of gravity while counteracting the tendency to slide out, while use of recline can make a huge functional difference in seating outcomes with respect to posture (see Pictures 4 and 5), as well as breathing and digestion. Using tilt and recline to alleviate negative influences of gravity on sitting posture can be done with manual and power wheelchairs.

- Manual frames—ultralight or lightweight adjustable frames can provide fixed tilt (seat slope) and recline (open hip angle) to benefit people with low tone. This is done through frame configuration and/or seating; for many people just a few degrees makes all the difference. This way a person can benefit from small degrees of tilt and recline with the transportability and light weight of a K0004 or K0005 frame.

• Manual tilt and recline frames (including adaptive strollers)—dependent wheelchair users have access to fixed and adjustable tilt-in-space, recline or combined mechanisms. Fixed tilt and recline can be configured through the frame and seating to limit options, if necessary. Self-tilting is an alternative for those who have the physical and mental ability to operate a switch (using a power tilt add-on) or self-tilt lever to change their own orientation.

• Power wheelchairs—use of power tilt and recline together in small increments of a few degrees can help low tone users find an ideal hip angle with adequate support to counteract both gravity and any tendency to slide out of the chair.

SEATING COMPONENTS

A wide variety of seating products work well for individuals with low tone once the correct mobility base and postural support needs are determined. Support surface contact with awareness and careful selection of cushion and back properties are used to stabilize the core—pelvis, back and upper legs. Secondary supports must be carefully selected. Pelvic positioning belt attachment points are crucial; if placed too far rearward (i.e. 45 degrees), some people with low tone will slide under and out, while others will collapse forward. Four-point pelvic belts offer better stabilization for some clients and can be configured to counteract anterior pelvic posture tendencies. Similarly, anterior trunk supports are chosen carefully for individuals with low tone who are "surface seekers" and tend to hang forward into the support. Back and trunk support selection, combined with use of tilt and recline can address this issue. Provide appropriate support for the individual's level of head control—this support may be needed full-time or only used when tilting/reclining or fatigued. Upper extremity supports like arm troughs or trays are used to assist with trunk positioning. These supports can prevent or significantly reduce upper limb drag, which negatively influences the upper body and head position, while increasing risk of potential injury to vulnerable joints.

SUMMARY

Clients with low tone have unique seating needs. Using the appropriate combination of seating and wheelchair configuration—based on comprehensive assessment findings inclusive not only of tone, but postural tendencies, daily activities, and prognosis for future change and comfort—will yield the best functional outcomes, promote participation and ultimately enhance the client's quality of life.

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