Wheelchair Provision

for Children and Adults
with Neuromuscular Conditions
in British Columbia

v1. April 2014

This manual was adapted with permission from the 2006 document entitled: *Wheelchair Provision for Children and Adults with Muscular Dystrophy and Other Neurological Conditions*, published in 2006 by the UK Muscular Dystrophy Campaign. The adaptations were undertaken by the ***British Columbia Wheelchair Guidelines Working Group*** to reflect the context, current practice and provision of wheelchairs for children and adults with neuromuscular conditions in British Columbia (BC). The adaptation process included appraisal, synthesis and integration of new research evidence; integration of the local perspectives and experiences of people with neuromuscular conditions and their service providers; and incorporation of the BC policy, funding and service environment.

Contact for the British Columbia Wheelchair Guidelines Working Group:

Sunny Hill Education Resource Centre
Sunny Hill Health Centre for Children
3644 Slocan Street, Room S225
Vancouver, BC V5M 3E8
Canada

Email: sherc@cw.bc.ca

This manual provides information about the needs of both children and adults with a neuromuscular condition. It is aimed at helping therapists plan appropriately with clients and families for wheelchair and seating provision. Although primarily focusing on therapists' roles, this manual may also be a useful resource for clients, parents, caregivers and other healthcare professionals.

The material in this manual and click-to-run evidence searches are available on the Child Development and Rehabilitation Website.

Members of the British Columbia Wheelchair Guidelines Working Group:

Sunny Hill Health Centre for Children

Roslyn Livingstone, *Occupational Therapist, Assistive Technology Team and Positioning & Mobility Team*

Maureen Story, *Physical Therapist, Positioning & Mobility Team*

Janice Evans, *Physical Therapist, Positioning & Mobility Team*

Lori Roxborough, *Occupational Therapist & Physical Therapist, Sunny Hill Evidence Centre*

University of British Columbia

Alison Hoens, *Physical Therapy Knowledge Broker*

Linda Del Fabro Smith, *Occupational Therapy & Occupational Science Knowledge Broker*

GF Strong Rehabilitation Centre

Cheryl Sheffield, *Occupational Therapist, Seating Services*

Cathy Husken, *Occupational Therapist, Seating Services*

Debbie Scott-Kerr, *Occupational Therapist, Outpatient Neuromuscularskeletal Program*

Shannon Sproule, *Physical Therapist, Outpatient Neuromuscularskeletal Program*

Ian Dennison, *Physical Therapist, Equipment Evaluation Program*

BC Children's Hospital

Lorelyn Meisner, *Physiotherapist, Neuromuscular Clinic*

Susan Garrett, *Occupational Therapist*

Cowichan Community School District 79

Susan Stacey, *Occupational Therapist*

Richmond Health Department

Sarah Pike, *Occupational Therapist, School Support Team*

Disability Health Research Network & International Collaboration on Repair Discoveries (ICORD)

Bonita Sawatzky, *Wheelchair Researcher*

The working group is immensely grateful for the extensive support of:

- **The UK Muscular Dystrophy Campaign** for permission to adapt their manual.
- **Clients and families** who reviewed documents and provided photos.
- **Charlotte Beck,** UBC Reference Librarian and **Andrea Ryce**, Sunny Hill Clinical Librarian for evidence search assistance.
- **The Canadian Agency for Drugs and Technology in Health (CADTH),** for the Rapid Evidence Response.
- **Mara Damin,** Sunny Hill Secretary, for management of working group documents.
- **Alyssa Barrie,** Occupational Therapist, for early content edits.
- **Veronica Naing,** Physical Therapist, for original illustrations.
- **Carolyn Andersson,** Clinical Education Officer, UBC Department of Physical Therapy, for all document formatting and final edits.

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Chapter 1 | Introduction

1.1 NEUROMUSCULAR ISSUES

This booklet covers all the muscular dystrophies as well as other groups of conditions that result in a similar type of muscle weakness, such as spinal muscular atrophy, various myopathies and myotonic disorders, and severe Charcot-Marie-Tooth disease. These disorders are often referred to collectively as 'neuromuscular conditions.'

See Appendix 1 for summaries of the main neuromuscular conditions.

Factsheets are available on most conditions from Muscular Dystrophy Canada.

Note that these conditions:

- Affect adults and children, males and females.
- Are genetic (more than one family member may be affected).
- Can cause muscle weakness, in both upper and lower limbs (proximal initially) and the trunk, neck and face.
- May cause weakness from birth or this may develop later.
- May mean that a child can never walk independently.
- Are progressive (at varying rates), therefore need regular review.
- Many run a predictable course.
- Cause deformities.
- Do not affect sensation (apart from Charcot-Marie-Tooth disease).
- Can mean that the person affected is unable to move independently.
- Can often cause respiratory muscle weakness.
- Can cause cardiomyopathy.
- Are currently incurable.
- Can cause abilities to fluctuate throughout the day, depending on activity levels and fatigue.
- Cause a risk of falls.
- Increase the risk of fractures, due to reduced bone density.
- Can cause function to be affected by changes in temperature.
- Can cause physical fatigue.

1.2 WHEELCHAIR NEEDS

Neuromuscular conditions progress at varying rates resulting in unique and changing needs

Some children with neuromuscular conditions will never be able to walk and will need to be provided with suitable wheelchairs and seating to enable them to develop independence and maximize their abilities. Information and resources on child development is available at the Child Development & Rehabilitation website.

For many people that are able to walk, a wheelchair will become essential at some stage. The rate of change will vary considerably from person to person, depending upon their particular condition and this will need to be taken into account when carrying out assessments and planning reviews. In most neuromuscular disorders, progression takes place at a steady rate. Generally, once ability is lost, it is very unlikely to be regained. Although

different conditions progress at different rates, in many cases, the pattern of weakness follows a predictable course.

Weakness of muscles around the hip girdle results in a "waddling" gait and difficulty walking distances, climbing steps, managing uneven surfaces and rising from sitting to standing. When walking, there is a tendency for the knees to "give way"; resulting in falls. Having fallen, there is extreme difficulty or, often, a complete inability to get up. There is a risk of fractures. Despite the fact they are still able to walk, people at this stage are likely to need assessment for their first wheelchair.

Accepting the need for a wheelchair affects people differently. It may be especially hard for parents of a child with a deteriorating condition. Be sensitive to these psychological issues while emphasizing that an appropriate wheelchair will help overcome fatigue and over-exertion, which may be harmful to muscles. Even if initial use is limited, having access to a wheelchair for days out, shopping trips etc. can help a client continue normal activities while gradually coming to terms with his or her changing mobility needs. Having a wheelchair can also prevent a 'crisis' situation if a fracture is sustained in a fall. The wheelchair will, inevitably, be needed more frequently. Delaying provision of an appropriate wheelchair at this time achieves little and is likely to cause a significant reduction in independence.

The walking ability of a person with a neuromuscular condition can vary from day to day, and even within a day, depending on activity levels. While he or she will be encouraged to walk for as long as possible, to maintain muscle strength, delay the onset of deformities and aid weight control, there comes a point when walking can only be considered therapeutic and is no longer functional. Individuals at this stage should be provided with an appropriate wheelchair to enable them to move around independently.

Many people will become dependent on a wheelchair and may need significant postural support. Because they cannot move themselves independently and are often sitting for long periods of time, they are at risk of developing pressure ulcers. Wheelchairs with specialist features can be invaluable for this group.

1.3 PLANNING AHEAD

It is essential to think and plan ahead - early referral should be encouraged. And as a provider you need to consider likely future needs. Both current and future wheelchair needs should be considered at each assessment.

People with neuromuscular conditions are generally reluctant to accept changes to seating. Encouraging compliance with using postural support at an early stage can help prevent future problems. It may be worth considering using products that can 'grow' or be adapted.

Children, particularly those with Duchenne muscular dystrophy, can rapidly deteriorate and this, combined with growth, creates constant change. Delays in assessment can cause deterioration in posture. Delays between assessment and provision may mean that a wheelchair and/or seating may no longer be suitable by the time it is provided. Any delay can be detrimental to the health and well-being of the individual.

When planning, remember the:

- Progression of the condition.
- Need to gain compliance with supportive seating.
- Need to avoid delays.

Consider:

- Future needs.
- Early provision of items that will be needed in future.
- Regular reviews.
- Adjustability in chosen products.

1.4 MULTIDISCIPLINARY TEAM

Community therapists provide seating services to both children and adults with neuromuscular conditions. Primary services for children are delivered through early intervention or school- based services. Specialty seating may be available in your region through child development centres, community health centre or regional hospitals. Provincial services for complex seating and mobility are available through Sunny Hill Health Centre for Children and GF Strong Rehabilitation Centre.

Check HealthLink BC for information on your local resources.

The GF Strong Rehab Centre *Assistive Technology & Seating Service* has assembled a list of questions to assist clients in choosing an equipment vendor. The team includes the client, vendor, therapist and the family.

1.5 OTHER ISSUES

It is vital that, when carrying out assessments, there is close liaison with all the professionals involved, as well as the client and his or her caregivers. Clients could be encouraged to inform all relevant professionals of a forthcoming assessment.

Consider:

- Daily living activities, particularly toileting.
- Transfers/lifts.
- Splints/orthoses.
- Feeding/swallowing difficulties.
- Other equipment e.g., mobile arm supports, ventilators.
- Medical issues e.g., spinal surgery, respiratory support.
- Vehicle/transport issues.

Although the role of the seating service is to assess for and provide appropriate wheelchairs and seating, during an assessment many clients will mention other areas of their lives with which they are having difficulty. It may be useful to highlight other services or equipment, or suggest they contact Muscular Dystrophy Canada for information.

Wheelchairs should enhance lifestyle and enable continued independence

Summary [1, 2]

Key issues to remember are:

- Neuromuscular conditions are mainly genetic with few if any treatments available. They cause progressive weakness but sensation is seldom affected.
- Mobility - "waddling gait," difficulty standing from sitting, frequent falls, many clients are unable to walk.
- Common features are: shoulder girdle weakness; poor head control; respiratory muscle weakness.
- Risk of contractures, deformities, scoliosis.
- Regular reviews are needed as changes in function and posture can happen quickly.
- Full assessment is needed. There are many issues to consider and liaison with other professionals is vital.

[1] Bushby K, Finkel R, Birnkrant DJ, Case LE, Clemens PR, Cripe L, et al. (2010). Diagnosis and management of Duchenne Muscular Dystrophy, part 2: Implementation of multidisciplinary care. *Lancet Neurol.* 2010; 9(2), 177-189.

[2] Wang CH, Finkel RS, Bertini ES, Schroth M, Simonds A, Wong B, et al. Consensus statement for standard of care in Spinal Muscular Atrophy. *J Child Neurol.* 2007; 22(8), 1027-1049.

Chapter 2 | Pre-school-aged Children

Neuromuscular conditions where a wheelchair is likely to be first needed in this age group are:

- Congenital muscular dystrophies.
- Congenital myopathies.
- Congenital myotonic dystrophy.
- (Severe) Charcot-Marie-Tooth disease (also known as Hereditary, Motor and Sensory Neuropathy).
- Spinal Muscular Atrophy (all childhood types).

INTRODUCTION

The first year of life is a period of rapid development from complete dependence to the emergence of independent sitting and mobility skills. Infants constantly interact with their environment through movement to develop and learn new skills. Toddlers begin to participate in daily living skills, gradually becoming more independent through the preschool years. They develop social skills; playing alone and with other children, both indoors and outdoors.

In contrast, children with neuromuscular conditions that affect early development, such as spinal muscular atrophy and congenital muscular dystrophy, have limited ability to move, to access their environment or to play with peers. Children with other neuromuscular conditions such as Duchenne muscular dystrophy may have a period of relatively normal development in this stage of life.

2.1 ABILITY AND NEEDS

Reduced self-locomotion in an infant and pre-school child as a result of neuromuscular conditions has a direct effect upon their cognitive and emotional development. Some will be unable to develop gross motor skills such as head control, bringing their hands to their mouth, sitting unaided, etc., because of low muscle tone. **Many will, however, still have the potential for normal intellectual (cognitive) development.**

Young children learn about people and objects through movement and interaction. They need to be active rather than passive recipients of experiences. Children who are unable to affect their environment through their own abilities can develop passive dependent behaviour where they give up trying to control their world. This can be well established by as early as four years of age.[1] It has been suggested that children who have limited abilities to move may pass a critical time for learning cognitive, emotional and social skills, if not provided with independent mobility experiences at an early age.[2]

An infant or pre-school child with a neuromuscular condition that affects early development will need assistance in gaining access to a stimulating environment where they can play and interact with their peers, care givers and toys. Some children may never walk, while others will walk independently for a limited amount of time. These children have restricted experiences and opportunities for play and exploration, and view their environment from floor level or while being held in the arms of their parents. This is obviously not the experience of their

[1] Butler C. Effects of powered mobility on self-initiated behaviours of very young children with locomotor disability. *Dev Med Child Neurol.* 1986; 28: 325.

[2] Tefft D, Guerette P, Furumasu J. Cognitive predictors of young children's readiness for powered mobility. *Dev Med Child Neurol.* 1999; 41: 665-70.

peers. All children of pre-school age need to test their environment and boundaries of behaviour. A child, who is not independently mobile, will not be aware that these boundaries exist. Children with physical disabilities often become frustrated at their functional limitations.

Many child development theories stress the importance of mobility in a child's development from a very early age. Independent mobility has been shown to play a critical role in the psycho-social development of children.[3] It is thought to be the physical, social, psychological and emotional experiences that are created by moving independently that contribute to the development of the personality along with visual-perceptual, language and cognitive skills.[4]

Independent movement is critical to normal development

A restriction in independent mobility could jeopardize intellectual and social development as well as well-being and happiness. Providing appropriate postural support and encouraging independent mobility helps overcome deficits in motor skills and allows psycho-social development to flourish.

Research has demonstrated that providing independent powered mobility to children as young as seven,[5] 14,[6] and 20 months[7] has helped them achieve developmental milestones, enhanced psychosocial skills and independence.

Early postural support and access to independent mobility can help children overcome many limitations and maximize their potential physical development.

> *"Once our daughter got her powered chair, she suddenly bloomed into this cheeky, inquisitive and playful two-year-old. She could keep up with her sisters and became the centre of attention. Her confidence just grew and grew."*
>
> **—Parent of a girl with SMA type II**

Initially postural supports can be added to the infant stroller, but, at the age when the child's peers are beginning to move and explore their environment, therapists should introduce alternative forms of mobility such as wheelchairs. Ultralight manual and power wheelchairs should be considered with appropriate postural supports.

Independent mobility helps develop initiative and enable personal choice. It is vital, therefore, to a small child's cognitive and psychosocial development that he or she can move around effectively and independently in his or her environment from an early age. As with any child, adult supervision at all times is imperative for their safety.

Factors to consider

- Independent movement is essential for cognitive and emotional development.
- Early provision of independent mobility may help prevent passive, dependent behaviour.
- Postural support should be provided early on within commercial baby equipment wherever possible.

[3] Kermoian R. Locomotion experience and psychological development in infancy. *In J Furumasu ed.* Pediatric powered mobility: Developmental perspectives, technical issues, clinical approaches. Arlington (VA); RESNA Press; 1997. P 7-22.

[4] Campos J J, Anderson D I, Barbu-Roth M A, Hubbard E M, Hertenstein M J, Witherington D. Travel broadens the mind. *Infancy*. 2000; 1(2): 149-219.

[5] Lynch A, Ryu J-C, Agrawal S, Galloway JC. Power mobility training for a 7-month-old infant with spina bifida. *Pediatr Phys Ther.* 2009; 21(4): 362-8.

[6] Jones MA, McEwen IR, Neas BR. Effects of power wheelchairs on the development and function of young children with severe motor Impairments. *Pediatr Phys Ther*. 2009; 24(2): 131-140.

[7] Jones MA, McEwen IR, Hansen L. Use of power mobility for a young child with spinal muscular atrophy. *Phys Ther.* 2003; 83: 253-62.

2.2 EMOTIONAL ISSUES

A toddler needs to develop basic autonomy, which leads to self-will. A young child with physical disabilities may be over-protected by anxious parents who constantly help him or her and, in doing so, may unintentionally reduce the opportunities to develop skills and reach full potential.

> "After a few doorframes were wrecked, he soon got the hang of it and now gets around with the skill of a rally driver. He's really happy now!"
>
> **—Father of a two-year-old with SMA type II**

The normal emotions of a child this age may be distorted or exaggerated. The curiosity of a toddler is boundless and his or her understanding of the world is self-centered.

As self-will develops so does emotional capacity: happy, sad, contented, angry etc. Each cognitive and motor milestone reached carries with it the development of emotion.

Normal social behaviours grow with emotional, cognitive and physical development. Providing a child with independent mobility complements emotional growth and maturity.

2.3 EQUIPMENT PROVISION

When considering this age group, there are three broad categories of children with neuromuscular conditions: non-sitters, sitters and walkers.[8] Their equipment needs are quite different and will be addressed separately.

Non-Sitters

These children are never able to be positioned in sitting without maximal support. They are diagnosed in early infancy with profound weakness and severe respiratory issues. Low tone, limited range of motion, poor head and poor postural control are apparent from an early age. Positioning considerations are mainly centred on maximizing respiratory function. Fatigue is a significant factor in this population and progressive scoliosis and chest wall changes may be apparent early. Swallowing and speech production may also be impaired. Assistive technologies should be considered to develop play skills and maximize participation.

Starting by six months of age, children should have customized postural supports in their infant stroller, high chair, car seat and crib. If their commercial baby equipment cannot be sufficiently modified, special needs strollers with options such as tilt and recline may be considered. Custom contoured seating may be required within this equipment. Many children in this group have a very limited life expectancy (below two years). Working within existing commercial equipment, or using recycled special needs strollers may be more appropriate than waiting long periods to order new equipment.

For children with less severe respiratory issues and an anticipated longer life expectancy, power mobility should be considered between 12 and 18 months. This has implications for funding of manual wheelchairs or strollers as a power chair is considered the primary device through At Home Program funding. Manual mobility equipment is then considered to be a 'back up' device (even though in reality manual and powered devices may need to be used equally to access different environments). The maximum amount available for manual mobility 'back up' equipment is $1500 if the child has a power mobility device.

Therapists may want to consider using the child's own stroller or using a recycled stroller through Red Cross Children's Medical Equipment Recycling and Loan Service (CMERLS) rather than using up funding by purchasing a new special needs stroller. If the child's needs require purchase of new manual and power mobility equipment,

[8] Wang CH, Finkel RS, Bertini ES, Schroth M, Simonds A, Wong B, Aloysius A, Morrison L, Main M, Crawford TO, Trela A, et al. Consensus statement for standard of care in Spinal Muscular Atrophy. *J Child Neurol.* 2007; 22(8): 1027-1049.

additional funding may be available through the Muscular Dystrophy Canada (MDC), extended health benefits, or other children's charities *(See Appendix 5).*

Equipment and Service Provision for Non-sitters

ISSUES	NEEDS	OUTCOMES
Assessment and Review	• Regular reassessments every 3-4 months.	• Early identification of issues. • Accommodate changing needs.
Postural Support	• Low tone – need full contact head, trunk and upper limb support. This may include contoured seating supports and either a tray or arm supports. • Need a variety of alternate positions in addition to sitting e.g. prone, supine, side-lying. • Equipment needs to adapt for child's growth. • Needs to be comfortable, 'breathable' and have extra covers.	• Minimize deformity. • Provide pressure redistribution if required. • Optimize respiratory function. • Maximize head control. • Maximize comfort.
Mobility Device	• Space and mounting for additional equipment e.g., suction, non-invasive ventilator, oxygen. • Allow for weight shift and change of position e.g., tilt and/or recline.	• Child can be moved around in different environments with all vital equipment. • Provide pressure redistribution if required. • Optimize respiratory function. • Maximize head control. • Maximizes comfort.
Aesthetics	• Acceptable to the family. • Need to see the child, not the equipment. • Soft, comfortable and child-friendly.	• Equipment will be used and allow participation in family and community life.
Transportation	• The safest place for young children is in a CSA-approved car seat. • Mobility devices should have tie down brackets so that they can be secured separately in the vehicle.	• Child is safely transported. • Encourage participation in family and community life.

Sitters

These children are diagnosed later in infancy and early childhood than the first group and often develop the ability to sit independently, although they may not be able to get into a sitting position by themselves. Some children may be unable to maintain sitting without some support, but all will be able to sit upright and maintain head control before they begin to lose abilities.

Key Issues | These children have low tone and are at high risk for developing a scoliosis. They need trunk support to help maintain postural alignment and, will often require head and upper limb support.

Between 6 and 12 months of age children should have customized postural supports in their infant stroller and high chair. Between 12 and 18 months mobility options either manual or power as appropriate should be considered.

Independent mobility is necessary for widening experiences, allowing safe exploration of the environment and participation with peers. Young children need mobility options that will allow them to keep up with peers and join in games. Children who can propel manual wheelchairs indoors should also be considered for power mobility outdoors. Young children need the opportunity to participate with peers independently.

There are many powered seat functions available: tilt; recline; elevating leg rests; elevation; lowering to the floor; and standing. Features such as standing have physiological benefits and standing, lowering to the floor and seat elevation allow increased access to the environment. However, standing and lowering to the floor are

not available on the same piece of equipment. Therapists and families need to prioritize needs and features due to funding restrictions and the limitations of wheelchair design.

In British Columbia, the At Home Program will typically fund a powered tilt system if it is medically justified. If a stand up feature is desired, *At Home Program* may fund $3,200 towards the addition of this feature using the funds from the alternate positioning budget. Additional funding will be required to fully fund this feature and to secure funding for any other options such as seat elevation, lowering to the floor, recline or elevating leg rests.

Some children will require both power and manual wheelchairs. If the child is able to self-propel the wheelchair for short distances then an ultralight manual wheelchair should be considered.[9] If the manual chair is mainly going to be moved by caregivers in order to allow the child to access environments where the power chair cannot be used due to access or transportation limitations, then a lightweight model will be adequate.

In either case, additional funding will be needed. In British Columbia a power chair is considered the primary device through *At Home Program* funding. Manual mobility equipment is then considered to be a 'back up' device. The maximum amount available for manual mobility 'back up' equipment is $1,500 if the child has a power mobility device. Therapists may want to consider using the child's own stroller or using a recycled manual wheelchair through *CMERLS* or seeking additional funding through sources such as *MDC* (*See Appendix 5*).

Equipment and Service Provision for Sitters

ISSUES	NEEDS	OUTCOMES
Assessment & Review	• Regular reassessments every 3-4 months.	• Permit early identification of issues. • Accommodate changing needs.
Postural Support	• Low tone – need head, trunk and upper limb support. • This may include planar or contoured seating supports and either a tray or arm supports. • A TLSO may be used as an adjunct to seating supports to help maintain spinal alignment. An abdominal cut-out in the TLSO is typically recommended to allow for diaphragmatic excursion. Care should be taken to monitor respiration in the TLSO and seating system. • Needs to be comfortable, 'breathable' and have extra covers. • Seating system needs to adapt for child's growth.	• Enable good posture with head and spinal/trunk support and pelvic alignment. • Minimize deformities. • Optimize respiratory function. Monitor respiratory pattern, respiratory rate, heart rate and oxygen saturation to assist in decision making when making changes to postural supports. • Provide pressure redistribution if required. • Maximize upper limb function and play. • Maximize visual stimulation. • Enable social interaction. • Enhance learning and emotional development.
Mobility Needs	• Adaptability is essential to accommodate the child in a number of positions e.g., tilt-in-space/recline, lying. • Seat elevation - may include to and from floor level. • Space and mounting for additional equipment. • Standing is an important goal at this age. Consider options that will allow child mobility in a standing position e.g., Go:Bot ; Standing Dani; sit to stand powered wheelchairs. • This age group is still very active in floor play and unless children are provided with specialized seating which lets them play at floor level they will be hindered.	• Enhance ability to play/interact with peers. • Enhance ability to learn play and exploration skills. • Able to be moved around in different environments with all vital equipment. • Able to access playground, rough outdoor terrain and keep up with peers over distance. • Able to move around at the same speed as their peers. • Able to participate in activities at different heights at home, preschool and community environments. • Able to use mobility device without fatiguing.

[9] Bushby K, Finkel R, Birnkrant DJ, Case LE, Clemens PR, Cripe L, et al. Diagnosis and management of Duchenne Muscular Dystrophy, part 2: Implementation of multidisciplinary care. *Lancet Neurol. 2010;* 9(2): 177-189.

ISSUES	NEEDS	OUTCOMES
Mobility Needs (continued)	• Joystick controls. Some children need to use both hands with the joystick positioned in midline. Some children are most successful with arm support angled towards midline and joystick mounted to arm support. Sensitive proportional or digital controls may need to be considered such as mini proportional joystick or track pad. • A lightweight manual wheelchair may be appropriate for some children in some environments e.g., indoors on smooth surfaces.	
Environmental Considerations	• To conform to the child's living and play environment. • Different functions for the child's different environments e.g., home may be unable to accommodate a power wheelchair – may need alternative seating system on a mobile base for indoors e.g., lightweight manual wheelchair. • If mobility device cannot lower to floor level, child may need alternate floor level seating system. • If mobility device does not have a stand up option, child may need a separate standing device – preferably one that allows independent mobility.	• Able to participate in activities at floor level, with peers and adults in a variety of environments.
Aesthetics	• Acceptable to family. • See child and not the equipment. • Self-esteem and equality with peers.	• Equipment will be used and enhance participation in community life. • Enhance child's awareness of body image/esteem, etc. • Empower child's confidence (choice of colour, etc.).
Transportation	• The safest place for young children is in a CSA approved car seat. • Mobility devices should have tie down brackets so that they can be secured separately in the vehicle.	• Child is safely transported. • Enhance participation in family and community life.

Ambulatory

These children are diagnosed later in childhood and are able to walk. They often have difficulty with transitions from floor, with stairs and are at risk for falls on uneven surfaces. In the preschool years, they may require either a power or manual wheelchair in order to keep up with peers, prevent falls outside and enhance participation.

Key Issues | Often families will be reluctant to consider a power wheelchair for a child who can walk indoors and may prefer to use a manual wheelchair outdoors as it is lighter and easier to transport. However, it is important to reinforce that this will not allow the child to develop age appropriate independence and to keep up with other children. Usually power mobility is necessary to enhance participation in the playground and in community environments.

In British Columbia a power chair is considered the primary device through *At Home Program* funding. Manual mobility equipment is then considered to be a 'back up' device. The maximum amount available for manual mobility 'back up' equipment is $1,500 if the child has a power mobility device. This group of children will benefit from having a lightweight or ultralight manual chair as well as a power chair. It may be possible to find a suitable recycled power or manual chair through CMERLS or to seek additional funding through sources such as MDC, extended benefits or children's charities *(See Appendix 5)*.

Equipment and Service Provision for Ambulatory Pre-school-aged Children

ISSUES	NEEDS	OUTCOMES
Assessment & Review	• Regular reassessments every six months.	• Enable early identification of issues. • Accommodate changing needs.
Postural Support	• Need firm seat and back with midline guides. • Seating system needs to adapt for child's growth.	• Enable good posture with trunk support and pelvic alignment. • Maximize function. • Minimize fatigue.
Mobility Device	• Lightweight manual chair may be needed if child can self-propel efficiently. • If parents are unable to transport a power wheelchair, a manual chair may be needed. • Power mobility may be needed to allow child to keep up with peers on playground and outdoors.	• Able to play/interact with peers. • Able to access playground, rough outdoor terrain and keep up with peers over distance. • Able to move around at the same speed as their peers. • Able to use mobility device without fatiguing.
Environmental Considerations	• To conform to the child's living and play environment.	• Child is able to keep up with peers and participate in a variety of environments.
Aesthetics	• Acceptable to family. • See child and not the equipment. • Self-esteem and equality with peers.	• Equipment will be used and enhance participation in community life. • Enhance child's awareness of body image/esteem, etc. • Empower child's confidence (choice of colour, etc.).
Transportation	• The safest place for young children is in a CSA approved car seat. • Mobility devices should have tie down brackets so that they can be secured separately in the vehicle.	• Child is safely transported. • Enhance participation in family and community life.

2.4 PLANNING AHEAD

Equipment management and funding considerations

When providing mobility equipment to young children it is important to be aware of the *At Home Program* funding limits and replacement periods. Up to date information can be found in the At Home Program Guide.

Currently the minimum replacement period for manual and power wheelchairs is five years. The minimum replacement period for a special needs stroller is three years. The young child's needs and environment may change considerably in this time so one needs to anticipate future needs and environmental considerations when ordering equipment. For example, when ordering a wheelchair for a two year old one needs to consider how that wheelchair will perform in the school environment and ensure that it will continue to allow the child to keep up with his peers as they grow and change.

One needs to consider that the child will grow considerably at this age and that the child's function will deteriorate over time in a progressive neuromuscular condition. Therefore, the equipment needs to be ordered with the necessary features to grow and to support the child as their function changes. When selecting the electronic package for the power wheelchair one needs to be certain that the electronics can support alternate access methods (e.g., a very sensitive joystick) if necessary.

Other issues such as how the equipment will be stored and transported need to be discussed before the equipment is ordered. Power wheelchairs need to be stored in a dry place protected from the weather and they need to be charged daily. Power wheelchairs weigh several hundred pounds and cannot be safely lifted into a vehicle. An adapted wheelchair accessible vehicle with tie downs to secure the wheelchair is recommended.

Safety and power mobility training

All young children require close supervision to keep them safe. Young children learning to use power wheelchairs are not like adults learning to drive a car, but are like other young children learning to walk. They have had no previous experience of moving independently and do not have well developed visual perceptual or spatial skills. They also lack emotional maturity and safety awareness.

Young children learning to walk are praised and encouraged for their efforts and allowed to learn at their own pace. They are not told to 'go in a straight line,' 'turn left,' 'be careful,' 'stop' etc.! Likewise, young children learning to use a power chair for the first time need to be allowed to learn by doing in a safe, play based environment.[10] The role of the adult is to encourage the child's skills through play and to keep them safe while they build skill.[11]

Adults can provide encouraging feedback but should remain quiet whenever possible to allow children time to figure out how to do things by themselves. Adults encouraging a child to play in the power chair should use only a few simple verbal suggestions such as 'come to mom' or 'let's go over there' rather than constant directional instructions such as 'go straight' and 'turn right' as this may suppress young children's learning.[12]

SUMMARY [1, 2]

Key issues to remember are:

- Emotional and psychosocial development of the child will be impacted by their experience of independent mobility.
- The physical, social and attitudinal environment will influence equipment choices.
- Planning ahead: life expectancy and anticipated rate of progression of muscle weakness will influence decisions.
- Postural support should be provided early on within commercial baby equipment wherever possible.
- Equipment management: when considering power mobility sheltered storage, charging facilities and transportation options should be discussed.
- Funding limitations for 'back up' manual mobility equipment should be considered when ordering power mobility equipment.

It is vital to a small child's cognitive and psychosocial development that, from an early age, they are able to mobilize effectively and safely in their environment

[10] Kangas K. *Powered mobility training for children with complex needs.* In: Proceedings of 18th International Seating Symposium, March 7-9, 2002. Vancouver BC. P 109-112.

[11] Durkin J. Discovering powered mobility skills with children: 'Responsive partners' in learning. *Int J Ther and Rehabil.* 2009; 16: 331-41.

[12] Wright-Ott C. The transitional powered mobility aid, a new concept and tool for early mobility. *In J Furumasu ed.* Pediatric powered mobility: Developmental perspectives, technical issues, clinical approaches. Arlington (VA): RESNA Press; 1997. P 58-69.

Chapter 3 | Schoolchildren to Adolescents

This chapter addresses the emerging and ongoing needs of school-aged children (ages 6-12) with neuromuscular conditions. 'Emerging' refers to the needs of children who are either newly diagnosed or losing their ability to walk. 'Ongoing' refers to the needs of children who are already wheelchair users, but whose needs may be changing.

Children with emerging needs:

- Congenital myopathies.
- Congenital myotonic dystrophy.
- Facioscapulohumeral muscular dystrophy (FSH).
- Limb girdle dystrophies.
- Duchenne muscular dystrophy.
- Severe Becker muscular dystrophy.
- Severe Charcot-Marie-Tooth disease (also known as Hereditary, Motor and Sensory Neuropathy).

Children with ongoing needs:

- Congenital muscular dystrophies.
- Congenital myopathies.
- Congenital myotonic dystrophy.
- Severe Charcot-Marie-Tooth disease.
- Spinal Muscular Atrophy (all childhood types).

INTRODUCTION

This age is a time of ongoing change for children as they move through childhood to adolescence. They are beginning to develop independence in many areas of everyday life, while consolidating early-learnt skills. At play and school, they start to undertake activities independently from their parents and other close adults. This is important as it helps them develop their individuality and build self-esteem. Children begin to learn right from wrong and the consequences of actions, both for themselves and others. There is so much to learn and it is a very exciting time for a child.

Children start to become interested in how they look, and want to be accepted by friends. They experiment with forming friendship groups and develop close relationships.

3.1 ABILITY AND NEEDS

During this period, many children will lose the ability to walk independently. A wheelchair plays a vital role in enabling the child to continue developing and learning alongside friends and family. Seating therapists have the ability to play a crucial part in promoting this process by assessing for and providing the appropriate wheelchair and seating system.

For many children with a neuromuscular condition, however, this is the time when they start to lose muscle strength and become more physically dependent. These children are coping with many changes to their bodies – the deterioration of physical skills and function – and becoming more aware of how they are different from others.

Factors to consider

- *Physical growth:* Children grow a lot between 5 and 12 years of age and mobility equipment should be ordered with potential for future growth.
- *Psycho-social development:* Children should be encouraged to become involved in activities with other children without adults always present.
- *Psychological readiness:* Accepting the need for mobility equipment and need for other technologies can be challenging for some children and families.
- *Transportation issues:* Mobility equipment should be ordered transport ready as children begin to travel in their mobility equipment in vehicles.

3.2 EQUIPMENT PROVISION

The equipment needs for children with emerging and ongoing needs are quite different and will be addressed separately.

Children with Emerging Needs

Some children, those with Duchenne muscular dystrophy for example, may still be walking independently. However, changes in motor skills will become more apparent as they get older and their muscles weaken.[1] [2] These children may tire and fatigue more than their peers and reduced muscle strength may cause problems with balance and frequent falling. Introducing the wheelchair for part-time use at this stage can prevent serious injury, such as fractures, which are more likely to occur in children with neuromuscular conditions as they have reduced bone density.[3] Ankle foot orthoses are often used at night to prevent development of contractures but are not recommended for use during the day as they make walking more difficult.[4]

> "When I got my powered chair, it was great. I could move about again and go to cubs with my friends; they all thought it was cool."
>
> **—Ben, 11 yr-old with DMD**

[1] Bakker JP, De Groot IJ, Beelen A, Lankhorst GJ. Predictive factors of cessation of ambulation in patients with Duchenne muscular dystrophy. *Am J Phys Med Rehabil.* 2002; 81: 906-912.

[2] Kohler M, Clarenbach CF, Bahler C, Brack T, Russi EW, Bloch KE. Disability and survival in Duchenne muscular dystrophy. *J Neurol, Neurosurg Psychiatr.* 2009; 80: 320-325.

[3] McDonald DG, Kinali M, Gallagher AC, Mercuri E, Muntoni F, Roper H, Pike MG. Fracture prevalence in Duchenne muscular dystrophy. *Dev Med Child Neurol.* 2002; 44: 695-698.

[4] Bushby K, Finkel R, Birnkrant DJ, Case LE, Clemens PR, Cripe L, Constantin C, et al. Diagnosis and management of Duchenne muscular dystrophy, part 2: Implementation of multidisciplinary care. *Lancet Neurol.* 2010; 9: 177-189.

Children may still have the ability to ambulate independently, but provision of a wheelchair allows them to choose how to spend their limited energy.[5] For example, a child may choose to use a wheelchair in the hallways at school to conserve energy and avoid being knocked down, but may park it at the door and still walk in the classroom. The loss of independent mobility is a challenging time for children, as they face the transition from walking to using a wheelchair. This is often extremely challenging as parents, siblings, extended family and friends, begin to adjust to life in a different way.

Introducing the idea of the first wheelchair raises issues for parents regarding not only the practical things such as access in the home environment and transport, but also the psychological issues around readiness and acceptance of the child's changing abilities. Many will have planned ahead but others must face the realization that changes will be needed at home. For example, in many older homes the doorways will not be wide enough to accommodate a wheelchair. Bedrooms being on different levels from the main entrance and small bathrooms may be other areas of consideration.

Many neuromuscular conditions follow a predictable course so it is possible to forecast how a condition will affect a child as he or she grows older. Working together with parents and local healthcare staff to establish an accurate diagnosis, if possible, will help wheelchair providers plan ahead to ensure the child's needs are met and appropriate funding resources explored. A referral to a seating therapist is important to best meet the child's and family's needs.

For children who are just beginning to use a wheelchair at this stage, it is important to proactively promote symmetrical posture through the use of firm support, rather than basic chair upholstery.[4] This practice begins when the child starts to need the wheelchair on a daily basis, even though he or she may still be able to stand and walk for short distances. When seated, the child is likely to be able to maintain an upright posture against gravity for short periods of time. However, when remaining in the position for longer periods, a kyphotic or asymmetric posture tends to develop because muscles prematurely fatigue. Postural support, such as a back support, trunk lateral support and contoured cushion, can help the individual return to a symmetrical posture.[6] The therapeutic goal is to provide appropriate support balanced with adequate freedom of movement, critical for the user to retain and maintain function.

Many children will be referred for a mobility device and there may be an assumption that a manual wheelchair comes first, to be followed down the road by a power wheelchair. However, experience has shown that these children are usually more independent in a power wheelchair and this should be considered as the first choice in an augmentative mobility device for most neuro-muscular conditions. For some children environmental or social considerations may make use of a power wheelchair difficult, in which case, a manual wheelchair may be the most appropriate choice. Children who have a power wheelchair will also require a manual wheelchair as a back-up for times when the power wheelchair is not available or for certain environments. Some children will begin using the power wheelchair only at school and have a manual wheelchair that their parents can transport easily for use at home.[5]

Consider the speed and range of the power wheelchair required to provide the user with independent outdoor mobility in a variety of environments and on a range of terrains. The basic wheelchair model may be insufficient to meet the current and changing needs of the child. When specifying a power wheelchair for children who are likely to experience increasing muscle weakness, it is important to anticipate features that may be needed within the five year replacement period such as power tilt and electronics that will support changing access needs. Ensure that the wheelchair is ordered with these features as it is much more expensive and impractical to add them on later.

[5] Mannlein J, Pangilinan PH. Wheelchair seating for children with Duchenne Muscular Dystrophy. *J Pediatr Rehabil Med.* 2008; 1: 225-235.

[6] Clark J, Michael S, Morrow M. Wheelchair postural support for young people with progressive neuromuscular disorders. *Int J Ther Rehabil.* 2004; 11: 365-371.

Participation in sports will also influence the types of wheelchair prescribed. For example: Power Soccer players often prefer rear wheel drive power wheelchairs and may need additional features added on such as fans to cool the motors which tend to overheat during an intense game.

Equipment and Service Provision for Children with Emergent Needs

ISSUES	NEEDS	OUTCOMES
Assessment & Review	• Regular reassessments every 6-12 months.	• Early identification of issues. • Accommodate changing needs.
Postural Support	• Need firm seat and back with midline guides. • Seating system and wheelchair need to adapt to the child's growth and changing needs. • Armrests set at appropriate height and width so that they provide upper extremity support without the client having to lean to one side.	• Enable good posture with trunk support and pelvic alignment. • Maximize function. • Minimize fatigue.
Mobility Needs	• Lightweight manual chair may be needed if child can self-propel. • Consider power wheelchair or power assist wheels. • Power mobility is often needed to access community and for longer distances. • A quadlink joystick mount will allow child to easily access desks, tables and work surfaces.[7] • Tray may be needed to provide appropriate work surface for school setting if unable to access suitable table.	• Prevent over-use injuries. • Enhance participation in home, educational, work and community environments. • Maintain functional skill. • Maintain independence throughout the day. • Reduce development of dependence or poor compensatory strategies.
Aesthetics	• Child is becoming more aware of body image and appearance. • Self-esteem and equality with peers.	• Empower child and increase confidence by involving in equipment and colour choices.
Environmental Considerations	• Equipment is suitable for the child's living, recreational, and educational environments. • More than one wheelchair may be required (e.g., manual wheelchair indoors, power wheelchair outdoors). • Equipment choices may be influenced by participation in sports. May access sports equipment through CMERLS or sports associations.	• Child is able to keep up with peers and participate fully in home, educational, leisure and community environments.
Equipment Management & Maintenance	• Understand how to access technical and maintenance support for mobility equipment. • Need for insurance coverage in case of loss or damage.	• Family is able to manage and maintain mobility equipment independently. • Ensures equipment is covered under home/school policy.
Transportation	• Consider how the wheelchair will be put in/taken out/transported in the vehicle. • Mobility devices should have tie down brackets so that they can be secured in a vehicle. • Wheelchairs should be ordered with headrests for transportation even if not required for postural support.	• Able to maintain access to vehicle and ability to travel. • Ensures method of transport meets approved safety standards. • Enhance participation in family, educational and community life.
Loss of Ability & Maintaining Independence	• Power chairs should be ordered with adaptable electronics to accommodate changes of driver interface if needed in future. • Power wheelchairs should be ordered with power tilt to accommodate loss of function. • Consider referral for counselling or involvement in peer support groups.	• Maintain independence despite loss of function. • Promote good posture and alignment. • Minimize pain and deformity.

[7] Evans S, Neophytou C, DeSouza L, Frank AO (2007). Young people's experiences using electric powered indoor-outdoor wheelchairs (EPIOCs): Potential for enhancing users' development? *Disabil Rehabil.* 2007; 29: 1281-1294.

Children with Ongoing Needs

Children, who were diagnosed with a neuromuscular condition during their early years, may already be using a wheelchair to maintain their mobility. Some have already been full-time power wheelchair users for a few years whereas others will be progressing from part-time use to full-time use of their wheelchair.

Key Issues

Some children will demonstrate increasing weakness in the upper limbs making it difficult to lift their arms against gravity. It may become more fatiguing to use the standard side mounted joystick. Some children will benefit from having the joystick mounted inboard in line with their shoulder. Angling the joystick towards midline so that a forward push is on an angle rather than straight forward may also be helpful as strength decreases.

As weakness increases, it becomes difficult to lift hands off the driver interface in order to reach switches used to operate auxiliary functions of the wheelchair such as turning it on and off or accessing speed or different modes. Integrating these functions through the joystick and reducing the number of switches is often helpful. Typically, switches need to be small, easy to activate and either positioned so they can be reached without moving the hand from the driver interface or accessed with the other hand.

Some children are able to control the driver interface only if their arms and hands are in a very specific position. Tilting the chair or moving over rough ground may cause them to lose this position and they are unable to reposition their arms independently. Use of larger armrest pads with elbow blocks, or arm troughs may help alleviate this problem.

If a new power wheelchair is being considered, it may be advisable to explore the use of a more sensitive joystick (e.g., mini or micro joystick) or alternate controls such as a touch pad to help maintain independence in driving in the future. Integrating computer access and other technologies such as EADLs (environmental controls) through the power wheelchair may be helpful. Referral for an assistive technology assessment may be recommended.

Children with conditions such as SMA type II may have developed significant postural asymmetries, including contractures, pelvic obliquity, and scoliosis. Children who are changing rapidly may require follow-up seating appointments every three to six months.[8] Seating systems for this group are often custom contoured to accommodate postural asymmetries and to optimize support and pressure distribution. Power tilt is often used to allow children to change position throughout the day.

See Chapters 6-9 for further information on assessment, seating and wheelchair features.

Spinal surgery may be considered at this stage to correct severe and increasing scoliosis. For children sometimes VEPTR (growing rods) are considered. This means that surgeries and seating changes will occur approximately every six months until a permanent spinal rod is inserted. The clients seating and functional abilities will change dramatically after surgery. Planning for the surgery needs to include plans to address the seating and mobility equipment needs post-surgery. Following spinal surgery, children will be transferred using a sling and lift system. Modifications to seating may be required to allow a sling to be put on and off with the child in the wheelchair (*See Appendix 2).*

[8] Richardson M, Frank AO. Electric powered wheelchairs for those with muscular dystrophy: Problems of posture, pain and deformity. *Disabil Rehabil. Assist Technol.* 2009; 4(3): 181-188.

Equipment and Service Provision for Children with Ongoing Needs

ISSUES	NEEDS	OUTCOMES
Assessment & Review	• Regular reassessments at least every 6 months. Clients with rapidly changing needs will need more regular review. • Children grow significantly between ages 6-12.	• Permit early identification of issues. • Accommodate changing needs.
Postural Support	• Low tone – need head, trunk and upper limb support. This may include planar or contoured seating supports and either a tray or preferably arm supports. • A TLSO may be used as an adjunct to seating supports to help maintain spinal alignment. An abdominal cut-out in the TLSO is typically recommended to allow for diaphragmatic excursion. Care should be taken to monitor respiration in the TLSO and seating system. • Headrests should have forward, backward and lateral adjustment as well as height adjustment. • Seating should be adaptable to client's growth. • Pressure relieving surfaces should be considered. • Power seating functions such as tilt-in-space and sit to stand should be considered. • Scoliosis is common in children with conditions such as SMA. Custom contoured seating may be necessary. • Following surgery, seating needs should be reassessed. • Flip down or swing away laterals may be needed to accommodate use of slings and lift system.	• Enable good posture with head and spinal/trunk support and pelvic alignment. • Ensures optimum support for upper limbs to improve posture and maximize upper limb and hand function. • Maximize participation in family, educational, and community life. • Minimize deformity. • Provide pressure redistribution if required. • Maximize head control. • Maximize comfort. • Maintain skin integrity. • Minimize pain. • Minimize fatigue. • Maximize upper limb function. • Maximize respiratory function.
Mobility Needs	• Adaptability to accommodate client in a number of positions e.g., tilt-in-space, and possibly standing if use of a separate standing frame is impractical. • Powered sit to stand features may enable the child to maintain independent standing without the need for transfers. • Driver interface: with increasing weakness, it becomes more difficult to use a standard proportional joystick. • Power wheelchairs should be ordered with programmable electronics as more sensitive proportional or digital controls may need to be considered.	• Assists with management of personal care. • Assists caregivers with moving and handling. • Allow child to change position throughout the day. • Extend duration of the ability to stand thereby helping to maintain bone density, prevent/delay the development of contractures, assist with kidney, bladder and gastro-intestinal function and enhance self-esteem. • Maintain independence in mobility throughout the day. • Minimize fatigue. • Reduce tendency to asymmetrical postures caused by positioning of driver interface or effort required for use. • Enhance participation in home, educational, and community environments.
Aesthetics	• Child is becoming more aware of body image and appearance. • Self-esteem and equality with peers.	• Empower child and increase confidence by involving in equipment and colour choices.
Environmental Considerations	• More than one wheelchair maybe required e.g., back-up manual wheelchair with tilt- in-space to access certain environments.[7] • Equipment is suitable for the client's living, recreational, and educational environments. • Consider home accessibility. • Consider speed and range of the wheelchair required to provide mobility in a variety of environments and terrains. • Power seat elevation may allow access to activities at varying heights.	• Enhance participation in home, educational, recreational and community environments. • Mobility equipment is suitable for participation in chosen sports e.g., Power Soccer.

ISSUES	NEEDS	OUTCOMES
Equipment Management & Maintenance	• Understand how to access technical and maintenance support for mobility equipment. • Need for insurance coverage in case of loss or damage.	• Family is able to manage and maintain mobility equipment independently. • Ensures equipment is covered under home/school policy.
Transportation	• Consider how the wheelchair will be put in/taken out/transported in the vehicle. • Mobility devices should have tie down brackets so that they can be secured separately in a vehicle. • Arm troughs are preferred over a tray for children who require arm support at all times as a tray will have to be removed during transportation.	• Able to maintain access to vehicle and ability to travel. • Enhance participation in family, educational and community life.
Loss of Ability & Maintaining Independence	• Power chairs should be ordered with adaptable electronics to accommodate changes of driver interface, integration of power seating functions through the driver interface and future integration of access to computer and environmental controls. • Different or repositioned switches may be required to operate auxiliary functions (turning chair on/off or changing modes) or for computer clicks.	• Maintain or increase independence despite loss of function. • Improve quality of life. • Promote good posture and alignment. • Minimize pain and deformity. • Optimize respiratory function.

3.3 PLANNING AHEAD

Equipment management and funding considerations

When providing mobility equipment to young children it is important to be aware of the *At Home Program* funding limits and replacement periods. Up to date information can be found in the At Home Program Guide.

> *"I felt much better once I had my wheelchair at school. I used it in the playground to begin with. I was happy because I could keep up with the others and I didn't need anyone with me in case I fell over."*
>
> **—Joe, 13 (Duchenne muscular dystrophy)**

Currently the minimum replacement period for manual and power wheelchairs is five years. The school child's needs and environment may change considerably in this time so one needs to anticipate future needs and environmental considerations when ordering equipment. One needs to consider that the child will grow considerably at this age and that the child's function will deteriorate over time in a progressive neuromuscular condition. Therefore, the equipment needs to be ordered with the necessary features to grow and to support the child as their function changes. When selecting the electronic package for the power wheelchair one needs to be certain that the electronics can support alternate access methods (e.g., a very sensitive joystick) if necessary.

Other issues such as how the equipment will be stored and transported need to be discussed before the equipment is ordered. Power wheelchairs need to be stored in a dry place protected from the weather and they need to be charged daily. Power wheelchairs weigh several hundred pounds and cannot be safely lifted into a vehicle. An adapted wheelchair accessible vehicle with tie downs to secure the wheelchair is recommended.

Given the often predictable, progressive nature of many of these conditions it is possible to plan ahead, and it is important to work to maintain the best possible sitting posture as loss of muscle strength occurs

See Chapters 6 - 9 for further information on seating, assessment and wheelchair features.

Chapter 4 | Adolescents to Young Adults

This chapter addresses the emerging and ongoing needs of adolescents and young adults with neuromuscular conditions. 'Emerging' refers to the needs of individuals who are either newly diagnosed or losing their ability to walk. 'Ongoing' refers to the needs of individuals who have had a diagnosis for several years and are already wheelchair users, but whose needs may be changing.

Clients with emerging needs:

- Becker muscular dystrophy.
- Facioscapulohumeral muscular dystrophy.
- Limb girdle muscular dystrophies.

Clients with ongoing needs:

- Becker muscular dystrophy (severe).
- Congenital muscular dystrophies.
- Congenital myopathies.
- Duchenne muscular dystrophy.
- Facioscapulohumeral muscular dystrophy.
- Limb girdle muscular dystrophies.
- Severe Charcot-Marie-Tooth disease.
- Spinal Muscular Atrophy.

INTRODUCTION

Adolescents and young adults become increasingly independent in all aspects of their lives. They go out and meet friends, form relationships, follow their interests and hobbies, participate in a range of activities and begin to think about future educational, employment and social opportunities.

Adolescents and young adults with neuromuscular conditions, like their peers without neuromuscular conditions, have a right to full participation in all aspects of society.

This should include:

- Maximized independence.
- Access to home environments.
- Access to educational environments.
- Access to work environment.
- Access to recreation and leisure.
- Access to transport.
- Being comfortable.
- Choosing how they look.
- Feeling empowered to make own life choices.
- Having positive self-esteem.

4.1 ABILITY AND NEEDS

In contrast to their peers, adolescents and young adults with progressive neuromuscular conditions are experiencing physical changes which may cause them to become more dependent on their parents, care givers and friends for their daily living, social and mobility needs. Progression of the condition, combined with growth, may result in constantly changing needs and abilities. As the condition progresses and mobility deteriorates, adolescents and young adults are faced with changes in functional ability, the need for a wheelchair, and being unable to keep up with peers or participate in activities they have previously enjoyed. Loss of independence for this age group can be a major area of concern and often causes feelings of isolation and/or depression. By this stage, adolescents and young adults with a neuromuscular condition may require a power wheelchair for indoor and outdoor mobility. The transition from walking to using a wheelchair can be emotionally and physically challenging for the client and his or her family and friends.

Once a physical ability is lost it is very unlikely to be regained. Timely implementation of appropriate supports and technology is needed to maximize function and participation.

Provision of an appropriate wheelchair and seating system is a critical factor in enabling and sustaining independence and participation

Looking and feeling good is important for this age group. Adolescents and young adults want to increase their independence from parents and make the most of their lives. This involves exploring the wider world by themselves as well as forming and maintaining relationships. They need to be able to socialise with friends, follow interests such as sport, music, shopping, movies, computers and electronic games, social networking, restaurants and clubs.

Factors to consider

- A rapid growth spurt during early teenage years can result in significant postural changes including development of contractures e.g., hip flexors, hip abductors, hamstrings, heel cords and also the development of scoliosis.
- Losing function and the increasing need for use of mobility equipment or additional technologies can be emotionally challenging for adolescents and young adults.
- Maintaining participation with peers is very important for this age group.

4.2 EQUIPMENT PROVISION

The equipment needs for adolescents with emerging and ongoing needs are quite different and will be addressed separately.

Adolescents with Emerging Needs

Adolescents and young adults with emerging needs will have been able to meet their mobility needs through walking until this age. However, due to increasing muscle weakness, they now need to use either a manual or power wheelchair for longer distances.

Key Issues

For clients who are just beginning to use a wheelchair at this stage, it is important to proactively promote symmetrical posture through the use of firm support, rather than basic chair upholstery.[1] This practice begins when the client starts to need the wheelchair on a daily basis, even though he or she may still be able to stand and walk for short distances. When seated, the client is likely to be able to maintain an upright posture against gravity for short periods of time. However, when remaining in the position for longer periods, a kyphotic asymmetric posture tends to develop because muscles prematurely fatigue. Postural support, such as a back support, trunk lateral support and contoured cushion, can help the individual return to a symmetrical posture.[2] The therapeutic goal is to provide appropriate support balanced with adequate freedom of movement, critical for the user to retain and maintain function.

Many clients will be referred for a mobility device and there may be an assumption that a manual wheelchair comes first, to be followed down the road by a power wheelchair. However, experience has shown that these clients are usually more independent in a power wheelchair and this should be considered as the first choice in an augmentative mobility device for most neuro-muscular conditions. For some clients environmental or social considerations may make use of a power wheelchair difficult, in which case, a manual wheelchair may be the most appropriate choice. Clients who have a power wheelchair will also require a manual wheelchair as a back-up for times when the power wheelchair is not available or for certain environments.

Some clients with conditions that are milder and have a later onset e.g., Becker muscular dystrophy, Facioscapulohumeral muscular dystrophy or some limb girdle dystrophies may use a lightweight manual wheelchair for long distances while they continue to walk in most environments. However, even clients with these conditions will be more independent in a power wheelchair and it is important to consider the functional needs of the client and the environments in which the wheelchair will be used.

Consider the speed and range of the power wheelchair required to provide the user with independent outdoor mobility in a variety of environments and on a range of terrains. In addition, the user, like any other young person, will want to look good and 'street credibility' will be important to him or her. The basic wheelchair model may be insufficient to meet the current and changing needs of the user.

When specifying a power wheelchair for clients who are likely to experience increasing muscle weakness, it is important to anticipate features that may be needed within the five year replacement period such as power seating functions and electronics that will support changing access needs. For most clients, a power tilt function should be included.

Additional features such as recline, powered elevating leg rests, seat elevation and powered stand-up functions should also be considered prior to ordering a first power wheelchair as it may be more expensive or difficult to add them later.

[1] Bushby K, Finkel R, Birnkrant DJ, Case LE, Clemens PR, Cripe L, Constantin C, et al. Diagnosis and management of Duchenne muscular dystrophy, part 2: Implementation of multidisciplinary care. *Lancet Neurol.* 2010; 9: 177-189.

[2] Clark J, Michael S, Morrow M. Wheelchair postural support for young people with progressive neuromuscular disorders. *Int J Ther Rehabil.* 2004; 11: 365-371.

Equipment and Service Provision for Adolescents with Emerging Needs

ISSUES	NEEDS	OUTCOMES
Assessment & Review	• Regular reassessments every 6-12 months.	• Early identification of issues. • Accommodate changing needs.
Postural Support	• Need firm seat and back with midline guides. • Seating system and wheelchair need to adapt to the client's growth and changing needs. • Armrests set at appropriate height and width so that they provide upper extremity support without the client having to lean to one side.	• Enable good posture with trunk support and pelvic alignment. • Maximize function. • Minimize fatigue.
Mobility Needs	• Lightweight manual chair may be needed if client can self-propel. • Consider power wheelchair or power assist wheels. • Power mobility is often needed to access community and for longer distances. • A quadlink joystick mount will allow client to easily access desks, tables and work surfaces.[3]	• Prevent over-use injuries. • Enhance participation in home, educational, work and community environments. • Maintain functional skill. • Maintain independence throughout the day. • Reduce development of dependence or poor compensatory strategies.
Aesthetics	• Acceptable to client • Self-esteem and equality with peers	• Empower user and increase confidence by involving in equipment choices.
Environmental Considerations	• Equipment is suitable for the client's living, recreational, work and educational environments. • More than one wheelchair may be required (e.g., manual wheelchair indoors, power wheelchair outdoors).	• Client is able to keep up with peers and participate fully in home, educational, work and community environments. • Equipment is suitable for participation in chosen sport (i.e., Power Soccer).
Equipment Management & Maintenance	• Understand how to access technical and maintenance support for mobility equipment.	• Client is able to manage and maintain mobility equipment independently.
Transportation	• Consider how the wheelchair will be put in/taken out/transported in the vehicle. • Mobility devices should have tie down brackets so that they can be secured separately in a vehicle. • Wheelchairs should be ordered with headrests for transportation even if not required for postural support.	• Able to maintain access to vehicle and ability to travel. • Enhance participation in family, work, educational and community life.
Loss of Ability & Maintaining Independence	• Power chairs should be ordered with adaptable electronics to accommodate changes of driver interface if needed in future. • Power wheelchairs should be ordered with power tilt to accommodate loss of function.	• Maintain independence despite loss of function. • Promote good posture and alignment. • Minimize pain and deformity.

Adolescents with Ongoing Needs

Adolescents and young adults in this group may have already been full-time power wheelchair users for many years whereas others will be progressing from part-time use to full-time use of their wheelchair

Key Issues | Adolescents and young adults with ongoing needs will demonstrate increasing weakness in the upper limbs making it difficult to lift their arms against gravity. It may become more fatiguing to use the standard side mounted joystick. Many adolescents will benefit from having the joystick mounted inboard in line

[3] Evans S, Neophytou C, DeSouza L, Frank AO (2007). Young people's experiences using electric powered indoor-outdoor wheelchairs (EPIOCs): Potential for enhancing users' development? *Disabil Rehabil.* 2007; 29: 1281-1294.

with their shoulder. Angling the joystick towards midline so that a forward push is on an angle rather than straight forward may also be helpful as strength decreases.

As weakness increases, it becomes difficult to lift hands off the driver interface in order to reach switches used to operate auxiliary functions of the wheelchair such as turning it on and off or accessing speed or different modes. Integrating these functions through the joystick and reducing the number of switches is often helpful. Typically, switches need to be small, easy to activate and either positioned so they can be reached without moving the hand from the driver interface or accessed with the other hand.

Some adolescents are able to control the driver interface only if their arms and hands are in a very specific position. Tilting the chair or moving over rough ground may cause them to lose this position and they are unable to reposition their arms independently. Use of larger armrest pads or arm troughs may help alleviate this problem.

If a new power wheelchair is being considered at this stage, it may be advisable to explore the use of a more sensitive joystick (e.g., mini or micro joystick) or alternate controls such as a touch pad to help maintain independence in driving in the future. Integrating computer access and other technologies such as EADLs (environmental controls) through the power wheelchair may be helpful. Referral for an assistive technology assessment and to Technology for Independent Living is recommended.

Clients who have been in wheelchairs for some time may have developed significant postural asymmetries, including contractures, pelvic obliquity, and scoliosis. During the adolescent growth spurt there may be a rapid progression of these postural asymmetries. Clients who are changing rapidly may require follow-up seating appointments every three to six months.[4]

Seating systems at this stage are often custom contoured to accommodate postural asymmetries and to optimize support and pressure distribution. Any changes made to the seating system to accommodate the changing body structure need to be made with the client's function in mind. Often the smallest change in position will impact the clients function. Changes to the clients seating that improve posture but make it harder to complete a functional task will not be accepted by the client.[5]

Many clients at this stage will be in their wheelchair for up to 15 hours a day. Spinal pain is common along with pain in the legs, hips and knees.[4] Power elevating leg rests, in combination with tilt and recline, may be helpful in allowing clients to change position frequently, thus alleviating pain associated with immobility.

Power recline can be used to assist with activities of daily living such as dressing or use of urinal[6] but may be impractical for clients who have molded backrests. Power elevating leg rests may help in managing circulation, and prevent lower extremity swelling. A power stand-up feature may improve function and independence as well as help to maintain bone density and muscle range, and assist with kidney, bladder and gastro-intestinal function. Contractures and pain in the feet are commonly seen and it may be difficult for some clients to tolerate wearing splints. Customizing the footrest supports may be a more appropriate option.

See Chapters 6-9 for further information on assessment, seating and wheelchair features.

[4] Richardson M, Frank AO. Electric powered wheelchairs for those with muscular dystrophy: Problems of posture, pain and deformity. *Disabil Rehabil. Assist Technol.* 2009; 4(3): 181-188.

[5] Liu M, Mindeo K, Hanayama K, Fujiwara T, Chino N. Practical problems and management of seating through the clinical stages of Duchenne's Muscular Dystrophy. *Arch Phys Med Rehabil.* 2003; 84: 818-824

[6] Lacoste, M, Weiss-Lambrou R, Allard M, Dansereau J. Powered tilt/recline systems: Why and how are they used? *Assist Technol.* 2003; 15: 58-68.

Spinal surgery may be considered at this stage to correct severe and increasing scoliosis. The clients seating and functional abilities will change dramatically after surgery. Planning for the surgery needs to include plans to address the seating and mobility equipment needs post-surgery (*See Appendix 2*).

As respiratory muscles weaken many clients will begin to require will require non-invasive positive pressure ventilation during the day. Occasionally, a person with a neuromuscular condition will be ventilated via a tracheostomy. Equipment such as ventilators, oxygen, or suction may need to be accommodated within the wheelchair (*See Appendix 2*).

Equipment and Service Provision for Adolescents with Ongoing Needs

ISSUES	NEEDS	OUTCOMES
Assessment & Review	• Regular reassessments at least every 6 months. • Clients with rapidly changing needs will need more regular review.	• Permit early identification of issues. • Accommodate changing needs.
Postural Support	• Low tone – need head, trunk and upper limb support. This may include planar or contoured seating supports and either a tray or preferably arm supports. • A thoracolumbosacral orthosis (TLSO) may be used as an adjunct to seating supports to help maintain spinal alignment. An abdominal cut-out in the TLSO is typically recommended to allow for diaphragmatic excursion. Care should be taken to monitor respiration in the TLSO and seating system. • Headrests should have forward, backward and lateral adjustment as well as height adjustment. • Seating should be adaptable to client's growth. • Pressure relieving surfaces should be considered. • Power seating functions such as tilt-in-space, recline and elevating leg rests should be included to allow clients to change position independently. Fatigue, pain and pressure are common issues with this population. • Variation in body size (thin vs. obese) is a feature of some neuromuscular conditions and their medical management. • Scoliosis is common in this age group. Custom contoured seating may be necessary. Following surgery, seating needs should be reassessed.	• Enable good posture with head and spinal/trunk support and pelvic alignment. • Ensures optimum support for upper limbs to improve posture and maximize upper limb and hand function. • Maximize participation in family, educational, work and community life. • Minimize deformity. • Provide pressure redistribution if required. • Optimize respiratory function. Monitor respiratory pattern, respiratory rate, heart rate and oxygen saturation to assist in decision making when making changes to postural supports. • Maximize head control. • Maximize comfort. • Maintain skin integrity. • Minimize pain. • Minimize fatigue. • Maximize upper limb function.
Mobility Needs	• Adaptability to accommodate client in a number of positions e.g., tilt-in-space, recline and possibly standing. • Even with bracing, in this age group, functional walking is often limited. Use of a standing frame may be impractical. Powered sit to stand features may enable the user to maintain independent standing without the need for transfers. • Driver interface: with increasing weakness, it becomes more difficult to use a standard proportional joystick. Clients may be more successful with the arm support angled towards midline and joystick mounted to the arm support. Power wheelchairs should be ordered with programmable electronics as more sensitive proportional or digital controls may need to be considered.	• Assists with management of personal care. • Assists caregivers with moving and handling. • Allow user to change position throughout the day. • Extend duration of the ability to stand thereby helping to maintain bone density, prevent/delay the development of contractures, assist with kidney, bladder and gastro-intestinal function and enhance self-esteem. • Maintain independence in mobility throughout the day. • Minimize fatigue. • Reduce tendency to asymmetrical postures caused by positioning of driver interface or effort required for use. • Enhance participation in home, educational, work and community environments.

ISSUES	NEEDS	OUTCOMES
Aesthetics	• Acceptable to client. • Self-esteem and equality with peers.	• Empower user and increase confidence by involving in equipment choices.
Environmental Considerations	• More than one wheelchair may be required e.g., back-up manual wheelchair with tilt- in-space to access certain environments.[3] • Equipment is suitable for the client's living, recreational, work and educational environments. • Consider speed and range of the wheelchair required to provide mobility in a variety of environments and terrains. • Power seat elevation may allow access to activities at varying heights.	• Enhance participation in home, educational, work and community environments.
Equipment Management & Maintenance	• Understand how to access technical and maintenance support for mobility equipment.	• Client is able to manage and maintain mobility equipment independently.
Transportation	• Consider how the wheelchair will be put in/taken out/transported in the vehicle. • Mobility devices should have tie down brackets so that they can be secured separately in a vehicle. • Arm troughs are preferred over a tray as this group requires arm support at all times and a tray will have to be removed during transportation.	• Able to maintain access to vehicle and ability to travel. • Enhance participation in family, work, educational and community life.
Loss of Ability & Maintaining Independence	• Power chairs should be ordered with adaptable electronics to accommodate changes of driver interface, integration of power seating functions through the driver interface and integration of access to computer and environmental controls. • Different or repositioned switches may be required to operate auxiliary functions (turning chair on/off or changing modes) or for computer clicks. • Some clients will require respiratory support such as BiPAP, ventilator, suction or oxygen. The wheelchair needs to accommodate this equipment.	• Maintain or increase independence despite loss of function. • Improve quality of life. • Promote good posture and alignment. • Minimize pain and deformity. • Optimize respiratory function.

4.3 PLANNING AHEAD

In British Columbia, equipment funded either by the At Home Program (children under 18 years who live at home or children in care under 19 years) or *Health Assistance Branch* (adults who are funded through the Ministry of Human Resources and Social Development) will typically have a minimum replacement period of five years. Power wheelchairs are considered the primary device and manual mobility equipment is then considered to be a 'back up' device. The maximum amount available for manual mobility 'back up' equipment is $1500 if the client has a power mobility device.

Some clients will benefit from having a lightweight or ultralight manual chair or a tilt-in-space manual chair as well as a power chair. It may be possible to find a suitable recycled power or manual chair through *CMERLS* or *MERLS* or to seek additional funding through sources such as MDC, extended benefits or charities *(See Appendix 5).*

The *At Home Program* and *Health Assistance Branch* typically fund a powered tilt system if it is medically justified. If a stand up feature is desired, *At Home Program* may fund $3200 towards the addition of this feature using the funds from the alternate positioning budget. Additional funding will be required to fully fund this feature and to secure funding for any other options such as seat elevation, recline or elevating leg-rests. *MDC*

may be able to assist with top-up funding for power seating functions. Extended benefits or other charities may also assist with funding.

Up-to-date information can be found in the At Home Program Guide.

Any wheelchair provided needs to be reliable and access to an efficient repair service is essential. The client must have an active role in the selection of the vendor. If a power wheelchair breaks down, the client can lose independence, access to work/education/leisure/sport and be unable to sit comfortably. Clients are responsible to ensure that their equipment is well maintained and serviced regularly. Insurance of mobility equipment should be explored through the home insurance policy.

See How to Choose an Equipment Dealer from GF Strong.

Safety has been raised as an important issue in this age group. Young people and their parents have suggested that additional training may be important to enhance safe access to the community.3

Driving

Some adolescents and young adults with neuromuscular conditions will be interested in learning to drive themselves. In British Columbia, they need to first study for and pass the written test. At that time they will be sent to a specialized driver-training program for assessment to determine what type of adaptations they will require.

See page 18 of Vancouver Coastal Health's *Community Resource Directory* for Resources for Driver Rehab and Drive Able Programs.

Appropriate and suitable equipment enables people with neuromuscular conditions to live an active teenage/young adult lifestyle, to be significantly more comfortable and to be as independent of caregivers as possible. This can make a major difference to their self-esteem, quality of life and level of independence.

Chapter 5 | Adults

NEUROMUSCULAR CONDITIONS

This chapter addresses the emerging and ongoing needs of adults with neuromuscular conditions. 'Emerging' refers to the needs of individuals who are either newly diagnosed or losing their ability to walk. 'Ongoing' refers to the needs of individuals who have had a diagnosis for several years, are already wheelchair users, but whose needs may be changing.

Clients with emerging needs:

- Becker muscular dystrophy.
- Facioscapulohumeral muscular dystrophy.
- Limb girdle muscular dystrophies.

Clients with ongoing needs:

- Becker muscular dystrophy (severe).
- Congenital muscular dystrophies.
- Congenital myopathies.
- Duchenne muscular dystrophy.
- Facioscapulohumeral muscular dystrophy.
- Limb girdle muscular dystrophies.
- Severe Charcot-Marie-Tooth disease.
- Spinal Muscular Atrophy.

INTRODUCTION

The development of independence and autonomy continues into adulthood. It involves the establishment of personal and professional life roles. Adults may participate in educational development, vocational pursuits, recreational activities, and the forming and maintenance of relationships.

Adults with neuromuscular conditions continue to have the right to participate in all aspects of society.

Adults are entitled to:

- Independence.
- An accessible home environment.
- An accessible work environment.
- A social life.
- Form/maintain relationships.
- Have children and/or care for children.
- Accessible transport.

5.1 ABILITY AND NEEDS

There are two broad categories of abilities and needs within this age group: Those with emerging mobility limitations and those with ongoing needs. The equipment requirements for each group are quite different and will be addressed separately. 'Emerging' refers to the needs of individuals who are either newly diagnosed or losing their ability to walk. 'Ongoing' refers to the needs of individuals who have had a diagnosis for several years, are already wheelchair users, but whose needs may be changing.

In contrast to their peers, adults with progressive neuromuscular conditions may be more dependent on parents, care givers and friends for their daily living, social and mobility needs. As the condition progresses and mobility deteriorates, adults are faced with changes in functional ability, the need for a wheelchair, and being unable to keep up with peers or participate in activities they have previously enjoyed. Loss of independence for this age group can be a major area of concern and often causes feelings of isolation and/or depression. Adults with ongoing needs may require a power wheelchair for indoor and outdoor mobility. For those with emerging needs, the transition from walking to using a wheelchair can be emotionally and physically challenging.

The walking ability of a person with a neuromuscular condition can vary from day to day, and even within a day, depending on activity levels. It becomes difficult for the affected individual to walk distances, climb steps, manage uneven surfaces and rise from sitting to standing. Although individuals are encouraged to walk for as long as possible there comes a point when walking can only be considered therapeutic and is no longer functional or dependable. Individuals at this stage should be provided with an appropriate wheelchair to enable them to move around independently and safely.

> *"The loss of function was gradual; it was not something that I noticed. When I look back I say "Wow, I've really changed."*
>
> **— David, aged 32, SMA**

The psychological impact of losing the ability to walk causes difficulty for many adults. Helping them stay as independent as possible can help to lessen this sense of loss. This can be achieved by providing a wheelchair that suits their environmental, social and transport needs. Quality of life is not negatively impacted with loss of physical function[1] as long as self- sufficiency is retained.[2] Some individuals with emerging limitations may have some postural deformities that will need to be addressed through appropriate seating support.[3] Others may have upper extremity weakness and the impact of this should be fully investigated when considering equipment prescription.[2]

Factors to consider

- Enabling/maintaining independence is often the primary focus of this age group.
- Reduced independent ambulation, fatigue, or progression from manual to power mobility use can have a psychological impact that needs to be acknowledged.
- Reduced upper extremity strength and function as well as increased difficulty with transfers are common and need to be considered with regards to equipment selection.
- Consider referral for supportive counselling or networking with peers who have similar functional capabilities.

[1] Kohler M, Clarenbach CF, Bahler C, Brack T, Russi EW, Bloch KE. Disability and survival in duchenne muscular dystrophy. *J Neurol, Neurosurg Psychiatr*. 2009; 80: 320-325.

[2] Pellegrini, N, Guillon B, Prigent H, et al. Optimization of power wheelchair control for patients with severe Duchenne Muscular Dystrophy. *Neuromusc Dis*, 2004; 14(5): 297-300.

[3] Richardson M, Frank AO. Electric powered wheelchairs for those with muscular dystrophy: Problems of posture, pain and deformity. *Disabil Rehabil. Assist Technol*. 2009; 4(3): 181-188.

5.2 EQUIPMENT PROVISION

The equipment needs for adults with emerging and ongoing needs are quite different and will be addressed separately.

Emerging mobility limitations

Adults with emerging needs will have been able to meet their mobility needs through walking until this age. However, due to increasing muscle weakness, they now need to use either a manual or power wheelchair for longer distances.

KEY ISSUES

Individuals with emerging mobility limitations may be having increasing difficulty with ambulation or propelling a manual wheelchair. This may have a significant psychological impact and thus needs to be handled sensitively.[4]

The transition from walking to using a wheelchair can also cause difficulties with access in the home, work or education environment. Adults need to consider the ongoing changes that may be required in the future to maintain their independence and accessibility while gradually coming to terms with their changing mobility needs.

Even if initial use is limited, having access to a wheelchair for longer distances can enable an adult to continue normal activities to prevent fatigue and over-exertion. If a fracture is sustained following a fall, having a wheelchair available could prevent a mobility 'crisis' situation from arising.[5] The wheelchair will, eventually, be needed more often. Earlier provision of an appropriate wheelchair can help reduce fatigue, decrease risk of injury and maintain independence.

A full and detailed assessment will be needed *(See Chapter 7)* as well as liaison with the client, caregivers and relevant professionals. There is expert consensus that the introduction of postural support into the client's wheelchair prescription should be a proactive process.[3] It should begin when clients start to need the wheelchair on a daily basis, even if they are still able to stand and walk for short distances. Planned reviews are recommended because the client's needs will change as their condition progresses. The adult should be encouraged to initiate professional consultation when it is required.

The mobility base should be matched to the functional needs of the client. Previous practice focused on using a manual chair for as long as possible. Current practice is moving towards providing a power chair earlier even if clients are still able to use a manual chair. This enables the client to maintain function, prevent overuse injuries and continue to participate in multiple environments.

> *"Getting a power wheelchair was a big change – I was now able to go out and spend time with my friends without needing one of them to volunteer to push me."*
>
> **— Kulvir, aged 37 years, DMD**

Provision of wheelchair that does not promote independent use such as an attendant controlled wheelchair, or a self-propelling wheelchair that cannot be used outside, is not recommended. Consideration may be given to high performance manual wheelchairs (*See Appendix 4*) or power assist kits (*See Chapter 9*), however, many users will need an indoor/outdoor powered wheelchair.

[4] Stevens M. Lower limb orthotic management of Duchenne Muscular Dystrophy: A literature review. *J Pros Orth.* 2006; 18(4): 111-119.

[5] McDonald DG, Kinali M, Gallagher AC, Mercuri E, Muntoni F, Roper H, Pike MG. Fracture prevalence in Duchenne Muscular Dystrophy. *Dev Med Child Neurol.* 2002; 44: 695-698.

Mobility equipment needs to be adaptable as needs of the user change. Infrequent wheelchair users should be encouraged to consider equipment that will allow for modifications as use increases and physical limitations progress. This may include upgrading electronics or size adjustments.

If a client uses a power wheelchair, it is often considered the primary device by the various funding programs. This often results in restrictions on funding for manual mobility systems. Manual mobility equipment is then considered to be a 'back up' device (even if manual and powered devices are used to access different environments). If the funder is the Ministry of Housing and Social Development, the maximum amount available for manual mobility 'back up' equipment is $1500 if the client has a power mobility device. If the client's needs require purchase of new manual and power mobility equipment, additional funding may be available through the *Muscular Dystrophy Canada* extended health benefits, or other registered charities *(See Appendix 5).*

At this stage the user may be able to maintain an upright posture against gravity for short periods while seated, but over time a kyphotic, asymmetric posture tends to develop as muscles prematurely fatigue. Those with postural deformities will also require appropriate corrective or accommodating seating. Provision of a contoured cushion and back support will enable them to return to a symmetrical position. Even if no postural deformities are evident, provision of some degree of contoured and supportive seating is recommended to provide comfort, reduce fatigue and maintain postural alignment.[6]

Consideration should be given for the goals and lifestyle of the user, family and caregivers when selecting seating and mobility equipment. A wheelchair with appropriate postural support and additional power functions, as needed by the client for his or her lifestyle, will help maintain independence and function. Static sitting (without postural shifts) in a wheelchair for several hours at a time may cause discomfort and puts the client at risk for skin integrity issues. Clients may be unable to independently change their sitting position or redistribute pressure. Powered seating functions such as tilt-in-space, recline and elevating leg rests can enable users to independently change their position and relieve pressure or discomfort.[7] Careful consideration should be given to the type of control, and its positioning, to ensure the functions can be operated independently by the client.[2] Referral to a seating specialist to identify the optimal combination of power positioning functions is recommended.

When considering powered seating functions power tilt is often the first to consider. Power tilt allows a user to position shift to redistribute pressure or to alter centre of gravity. Backrest recline may also be considered to allow a client to change seat to back angle. Use of recline has been found to be beneficial for back spasm relief and may assist with personal care activities.

It is not advisable to use backrest recline without use of tilt. Doing so may result in: increased lordosis, forward sliding pelvis and shearing. Shearing forces may result in skin breakdown. To prevent these issues, it is recommended that the client tilt prior to recline.

[6] Liu M, Mindeo K, Hanayama K, Fujiwara T, Chino N. Practical problems and management of seating through the clinical stages of Duchenne's Muscular Dystrophy. *Arch Phys Med Rehabil*. 2003; 84: 818-824.

[7] Dicianno BE, Arva J, Lieberman JM, Schmeler MR, Souza A, Phillips K, et al. RESNA position on the application of tilt, recline and elevating legrests for wheelchairs. *Assist Technol*. 2009; 21(1): 13-22.

Difficulties with transfers and reaching may be overcome by the use of a powered wheelchair with seat elevation.[8] A power stand-up feature may improve function and independence as well as help to maintain bone density and muscle range, and assist with kidney, bladder and gastro-intestinal function.[9]

There are usually funding restrictions on the type of powered seating functions as each adds significant cost to the cost of the wheelchair system. Power tilt is generally funded but the other powered seating components usually require significant justification and may be considered by the funder to be beyond basic provision levels. If the wheelchair user is working or interested in attaining employment, they may be able to procure funding for extra wheelchair functions needed for his or her job through an alternative funding source (e.g., Employment Program for Persons with Disability [EPPD program], MDC, or EATi. *(See Appendix 5).*

Consider the client's vehicle and whether alterations are needed. If possible, it is recommended that the wheelchair user transfer out of the wheelchair when in a vehicle. If the wheelchair user remains in the wheelchair, the wheelchair should be equipped with four securement points and be used with a four-point shoulder strap tie down system. The safest position for the wheelchair is forward facing in the vehicle. Specialist information and advice is available from a driver rehab centre experienced in assessment for adaptive driving and modifying vehicles to accommodate wheelchairs. See page 18 of Vancouver Coastal Health's *Community Resource Directory* for Resources for Driver Rehab and Drive Able Programs.

See Chapters 6-9 for further information on assessment, seating and wheelchair features.

Equipment and Service Provision for Adults with Emerging Needs

ISSUES	NEEDS	OUTCOMES
Assessment & Review	• Assessment and consideration of roles, environments and client wishes. • Full assessment of ability to self-propel on a variety of surfaces and slopes/ramps. • Early introduction or discussion of potential adaptive equipment to maximize independence • More than one wheelchair may be required (e.g., manual wheelchair indoors/ power wheelchair outdoors). • Liaison or referral to other appropriate services such as recreation therapy or adaptive sport groups.	• Permit early identification of issues. • Accommodate changing needs. • Enable user to maintain independence or access to environments. • Enable user to participate in daily activities. • Client linked with appropriate services (Recreation Therapy, Counselling, Peer Support, Adaptive Sports).
Postural Support	• Appropriate supportive seating will promote good postural alignment. • Consideration of the weight of the postural seating components especially in manual wheelchairs. • Supportive seating requirements may be influenced by specific functional capabilities of the client. • Seating system needs to be comfortable. • Consideration should be given to various orthotic devices clients may use (i.e., abdominal binders, AFO, TLSO). • Seating system will ideally allow for minor weight fluctuations.	• Encourage maximal functional ability. • Maintain comfort. • Minimize development and severity of postural deformities. • Reduce risk of pressure problems. • Enable social participation.

[8] Arva J, Schmeler MR, Lange ML, Lipka DD, Rosen LE. RESNA position on the application of seat-elevating devices for wheelchair users. *Assist Technol*. 2009; 21(2): 69-72.

[9] Arva J, Paleg G, Lange M, Lieberman J, Schmeler M, Dicianno B, Babinec M, Rosen L. RESNA position on the application of wheelchair standing devices. *Assist Technol.* 2009; 21: 161-168.

ISSUES	NEEDS	OUTCOMES
Mobility Device	• Introduction of a power wheelchair as an adjunct to manual wheelchair use. • Provision of appropriate wheelchair, possibly with 'non-standard' features. • Consideration of transport and driving issues. • Early introduction or discussion of potential. adaptive equipment to maximize independence • Consider an ultra-lightweight manual wheelchair or power assist wheels. • Wheelchair features assist with transfers (e.g., Tilt, seat elevator, hand holds, higher seat to floor height, full length armrests).	• Promote user independence and encourages ongoing participation in multiple environments. • Prevent overuse injuries. • Encourage acceptance of assistive devices. • Reduce development of dependency or poor compensatory strategies. • Optimize prescription of transfer aids or equipment.
Environmental Considerations	• Consider assessment of user's home, school, work, and recreational/leisure environments. • More than one wheelchair may be required (e.g., manual wheelchair indoors/ power wheelchair outdoors). • Consideration of adaptive equipment such as mobile arm supports, page turners, computer access devices. • Liaison with other appropriate services such as recreation therapy or adaptive sport groups.	• Enable user to maintain independent mobility in all environments. • Promote independence in ADLs.
Transportation	• Consideration of how wheelchair will be put in/taken out/carried in the car. • Consideration of referral to driver rehab centre for assessment of adaptive controls and/or vehicle access equipment.	• Enable user to maintain access to vehicle and ability to travel. • Enable user to maintain access to vehicle and ability to travel.

ONGOING MOBILITY LIMITATIONS

Adults in this group may have already been full-time power wheelchair users for many years whereas others will be progressing from part-time use to full-time use of their wheelchair. Adults who have a neuromuscular condition need ongoing reviews of their equipment and care needs. They may now wish to take a more active role in directing their care and managing their medical concerns. Funding sources may change dependent on age, employment and financial status.

By the time they reach adulthood, most of this group will have very little active movement and function will be severely limited. They are also likely to have significant postural deformities. They may use electronic aids to daily living and other adaptive devices to assist with independence. Many will be using non-invasive ventilation to assist with breathing at night *(See Appendix 2*) and some may also need it during the day.[1] Some of these adults will have problems with circulation and be prone to fatigue. All these factors need to be considered when assessing for suitable wheelchairs and seating.

KEY ISSUES

- These clients often have significant postural deformities with very limited motor control of their fingers or heads.
- The independence of adults with neuromuscular conditions is intricately dependent on the reliability and function of their equipment. They often have alternate power mobility drive controls, powered seating function, electronic aids to daily living and they may need to carry a ventilator or suctioning machine on their wheelchair for daytime use.

Full and detailed assessments are recommended as needs are identified. Adults with neuromuscular conditions assume increased responsibility and should take a more proactive role in managing their mobility and equipment needs. This will include involvement from caregivers, other health professionals and medical equipment vendors. Adults should be encouraged to initiate professional consultation as needs change.

> "As the changes come I figure out ways to deal with it – to make it easier and to adapt. Technology is a big deal for me as it allows me to do many things independently."
>
> **— Sital, aged 41, DMD**

It is essential that the seating position can be changed to help redistribute pressure as this will improve comfort and enable independence. Promotion of a symmetrical and comfortable seating posture is attained through the use of firm and contoured support.[6] Consideration must be given to the functional implications of seating changes related to compensatory movements that individuals use. Supportive, pressure distributing seating along with powered seating functions allow the user to independently self-reposition.

Consideration should be given for the goals and lifestyle of the user, family and caregivers when selecting seating and mobility equipment. A wheelchair with appropriate postural support and additional power functions, as needed by the user for his or her lifestyle, will help maintain independence and function. Static sitting (without postural shifts) in a wheelchair for several hours at a time may cause discomfort and puts the client at risk for skin integrity issues. Users may be unable to independently change their sitting position or redistribute pressure. Powered seating functions such as tilt-in-space, recline and elevating leg rests can enable users to independently change their position and relieve pressure or discomfort.[7] Careful consideration should be given to the type of control, and its positioning, to ensure the functions can be operated independently by the user.[2] Referral to a seating specialist to identify the optimal combination of power positioning functions is recommended.

When considering powered seating functions power tilt is often the first to consider. Power tilt allows a user to position shift to redistribute pressure or to alter centre of gravity. Backrest recline may also be considered to allow a user to change seat to back angle. Use of recline has been found to be beneficial for back spasm relief and may assist with personal care activities.

It is not advisable to use backrest recline without use of tilt. Doing so may result in: increased lordosis, forward sliding pelvis and shearing. Shearing forces may result in skin breakdown. To prevent these issues, it is recommended that the user tilt prior to recline.

Difficulties with transfers and reaching may be overcome by the use of a powered wheelchair with seat elevation.[8] A power stand-up feature may improve function and independence as well as help to maintain bone density and muscle range, and assist with kidney, bladder and gastro-intestinal function.[9]

There are usually funding restrictions on the type of powered seating functions as each adds significant cost to the cost of the wheelchair system. Power tilt is generally funded but the other powered seating components usually require significant justification and may be considered by the funder to be beyond basic provision levels. If the wheelchair user is working or interested in attaining employment, they may be able to procure funding for extra wheelchair functions needed for his or her job through an alternative funding source (e.g., Employment Program for Persons with Disability [EPPD program], MDC, or EATi. (*See Appendix 5*).

Transfers in and out of a vehicle may be difficult and some users may opt to travel in their wheelchair. A proper restraints system needs to be in place both on the wheelchair and in the vehicle. The wheelchair should be equipped with four tie-down brackets. It is suggested that a headrest system be used if the user sits in the wheelchair when in transit to protect from whiplash injury in the effect of an accident. Wheelchair trays and communication systems should be removed from the wheelchair when in transit. Specialist information and advice is available from a driver rehab centre experienced in assessment for adaptive driving and modifying vehicles to accommodate wheelchairs. See page 18 of Vancouver Coastal Health's *Community Resource Directory* for Resources for Driver Rehab and Drive Able Programs.

See Chapters 6-9 for further information on assessment, seating and wheelchair features.

Equipment and Service Provision for Adults with Ongoing Needs

ISSUES	NEEDS	OUTCOMES
Assessment & Review	• Assessment and consideration of roles, environments and client wishes. • Early introduction or discussion of potential adaptive equipment to maximize independence • More than one wheelchair may be required (e.g., Back up tilt in space manual wheelchair). • Liaison with or referral to other appropriate services such as Assistive Technology or Peer Mentors.	• Permit early identification of issues. • Accommodate changing needs. • Enable user to maintain independence or access to environments. • Enable user to participate in daily activities • Client linked with appropriate services (Assistive Technology, Counselling, Peer Support, PROP).
Postural Support	• Appropriate supportive seating will promote good postural alignment. • Supportive seating requirements may be influenced by specific functional capabilities of the client. • Consideration should be given to various orthotic devices clients may use (i.e., abdominal binders, AFO, TLSO). • Seating system will ideally allow for minor weight fluctuations. • Assessment for pressure risks and possible areas of concern: ischial tuberosities, coccyx, elbows, ankles, feet, scapulas. • Use of a powered tilt system is warranted to allow independent pressure redistribution, fatigue management or pain control. • Clients may have very limited head control and require a specialized headrest.	• Encourage maximal functional ability. • Minimize development and severity of postural deformities and risk of pressure problems. • Enable social participation. • Give user ability to change position independently to maintain comfort and a sense of stability or security. • Enable a user to lie back with head well supported for positional shifts or rests • Provide user with independence on ramps and slopes. • Reduce risks of falling forward from wheelchair. • Reduce need for restrictive harnesses/ straps etc. • Assist caregivers with moving and handling as well as the management of personal care.
Mobility Needs	• Adequate access to desks, tables and work surfaces (quadlink). • Independence in moving wheelchair and controlling seating features. • The wheelchair is an extension of the user's physical appearance and personal preferences regarding aesthetics need to be respected. • Increased weakness in upper limb/hand function may make it difficult to use standard joystick controls. Alternatives such as a light touch joystick may need to be assessed. • Expanded electronics will be required to accommodate alternate controls, and may be required for the integration of power seating features. • Different or repositioned switches may be required to change speed or function or these features may be integrated in the wheelchair electronics.	• Maintain functional skills. • Ensure user does not have to lean towards or away from control to obtain optimum leverage. • Maintain independence throughout the day. • User can move independently with all required equipment in different environments. • Maintain independence. • Improves quality of life.

ISSUES	NEEDS	OUTCOMES
Mobility Needs (continued)	• Some adults may require equipment such as ventilator, suction and oxygen. There needs to be space within the seating/wheelchair to accommodate this additional equipment.	
Environmental Considerations	• Support for upper limbs essential due to increasing upper limb weakness. • Different activities and it may require different positions. • User may be assessed for an environmental aid to daily living system to use in conjunction with wheelchair. • Consider the speed and range of the wheelchair required to provide the user with independent outdoor mobility in a variety of environments and on a range of terrains.	• Ensure optimum support for user's arms. • Maximize upper limb and hand function. • Maintain independence. • Contributes to overall postural alignment and comfort. • Improve quality of life.

5.3 PLANNING AHEAD

Mobility equipment needs to be adaptable as needs of the user change. This may include upgrading electronics or size adjustments. Funders often have time limitations before consideration will be given to purchase of a new mobility system. Currently clients who receive equipment funding from the Ministry of Human Resources and Social Development are only eligible for new equipment at minimum every five years. If a client uses a power chair, it is considered the primary device through the Ministry funding program. Manual mobility equipment is then considered to be a 'back up' device and the maximum amount available for manual mobility 'back up' equipment is $1500. Additional funding may be available through the *Muscular Dystrophy Association (MDC)*, extended health benefits, or other registered charities *(See Appendix 5)*.

Any wheelchair provided needs to be reliable and access to an efficient repair service is essential. The client must have an active role in the selection of the vendor. See Equipment Evaluation Services - How to Choose an Equipment Dealer from GF Strong. If a power wheelchair breaks down, the user can lose independence, access to education/work/leisure/sport and be unable to sit comfortably. Clients are responsible to ensuring that their equipment is well maintained and serviced when required. Insurance of the mobility device may need to be explored through the home insurance policy. The basic wheelchair model may be insufficient to meet the current and changing needs of the user.

SPECIAL CIRCUMSTANCES

During pregnancy, women with neuromuscular conditions who are usually able to walk may find this more difficult or impossible and may need access to a wheelchair. Full-time wheelchair users may need modifications or an alternative wheelchair during pregnancy to accommodate growth. Functional independence may be impacted during pregnancy due to joint laxity or positional shifts. Centre of balance will likely be impacted and this should be considered in the set-up of the wheelchair and for transfers.

Many adaptations can be made to the wheelchair and the environment to assist the user to participate in parenting tasks such as feeding and carrying the baby. See Through the Looking Glass for useful resources.

SUMMARY:

- Seating and mobility equipment should support and enhance participation in home, educational, work and community environments.
- Postural support should be provided early on in the first wheelchair, even if the client is still able to walk short distances.
- Need to balance amount of support with the freedom of movement needed for function.
- Power wheelchairs for full-time wheelchair users with neuromuscular conditions are usually ordered with tilt. Power recline and elevating leg-rests should also be considered. Power seat elevation may assist with functional activities and participation.
- Power wheelchairs for all clients should be ordered with expandable electronics to allow future changes of driver interface and integration of power seating functions, environmental controls and access to other technologies through the power wheelchair.
- Funding limitations for 'back up' manual mobility equipment, power seating functions and integration of other technologies should be considered when ordering power mobility equipment.
- Several appointments may be needed to deliver and set up equipment optimally.
- Small adjustments may significantly impact independence.
- Equipment prescriptions need to be individually tailored to the lifestyle wishes of the client and his or her family.

Appropriate and suitable equipment enables people with neuromuscular conditions to live an active lifestyle, to be significantly more comfortable and as independent of caregivers as possible. This can make a major difference to their self-esteem, quality of life and level of independence.

Provision of appropriate wheelchairs and supportive seating at the correct time can enable adults with neuromuscular conditions to live full and active lives and decrease their dependence on others.

> *"I have tried to live as normal a life as possible and that of course requires using adaptations… Of course there are some limitations but that has not prevented me from doing what I want to do."*
>
> **— Kulvir, aged 37, DMD**

Chapter 6 | Principles of Seating

INTRODUCTION

Children and adults with progressive neuromuscular disorders may have very specific seating needs and meeting those needs can be challenging for wheelchair seating services.

Wheelchairs were originally designed to transport people from one place to another and now they have evolved to rank among the most important therapeutic and rehabilitative devices.[1] As technology has advanced, the expectations of many clients have risen and the gap between what is available and what is prescribed can often be frustrating. Applying for a first wheelchair can be an emotionally challenging time for a client as they may feel they have reached a significant milestone in the progression of the disease.[2, 3]

Coping with a condition where the goal posts are constantly moving is not easy and there is a degree of evidence that explores the psychosocial impact of progressive neuromuscular disorders upon families.[4] The progression of neuromuscular conditions can make it difficult to prescribe appropriate seating as frequent appointments may be required. It is necessary to ensure a review system is in place to be certain that the prescription delivered in previous months is still appropriate throughout the changing physical parameters of the condition.[5]

Twenty-four hour postural management is now an accepted approach for health professionals working within the field of neurodisability.[3] [6] Postural management is defined as 'the use of any technique to minimize postural abnormality and enhance function.' Therapists assess sitting, lying and standing postures to ensure an individual is able to adopt as symmetrical a posture as possible during prolonged periods of immobility while still being comfortable, able to function and maintain pressure redistribution techniques. When prescribing any form of postural support it is helpful if the therapist has a sound knowledge of the way the condition progresses in order to facilitate appropriate proactive intervention.[5]

This chapter explores fundamental aspects of appropriate seating and wheelchair prescription.

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[1] Gavin–Dreschnack D. Effects of wheelchair posture on user safety. *Rehabilitation Nursing.* 2004; 29 (6): 221-226.

[2] Farley R, Clark J, Davidson C, Evans G, MacLennan K, Michael S, Morrow M, Thorpe S. What is the evidence for the effectiveness of postural management? *International Journal of Therapy and Rehabilitation.* 2003; 10 (10): 449-455.

[3] Morrow M. Duchenne muscular dystrophy – A biopsychosocial approach. *Physiotherapy.* 2004; 90 (3): 145-150.

[4] Bushby et al. Diagnosis and management of Duchenne muscular dystrophy, Part 1 & 2. *Lancet Neurol*. Published online Nov 30, 2009. http://www.treat-nmd.eu/userfiles/The_diagnosis_and_management_of_DMD_Lancet_complete_with_erratum.pdf

[5] Poutney TE, Mulcahey CM, Clarke SM, Green EM. The Chailey Approach to Postural Management. 2004; Second edition Active Design, Birmingham.

[6] Bushby K, Bourke J, Bullock R, Eagle M, Gibson M, Quinby J. The multidisciplinary management of Duchenne muscular dystrophy. *Current Paediatrics*. 2005; 15: 292-300.

6.1 OPTIMUM SEATING POSITION

It is generally accepted that there are four main aims that may be considered when assessing an individual's seating and wheelchair:

- Achieve the optimal postural support to minimize developing deformity, taking account of the effects of gravity.
- Maintain/enhance functional ability.
- Promote comfort and provide appropriate support, which may vary between activities and environments throughout the day.
- Achieve client and family goals for participation in life roles.

Sharing information with clients and family members is fundamental to developing an understanding of the challenges that each user will face on a daily basis and will assist the therapist when prescribing specific equipment.[7]

6.2 INTRODUCTION OF POSTURAL SUPPORT

There is currently no definitive research to define the correct time at which postural support should be introduced into the wheelchair prescription of someone with a neuromuscular condition. Expert opinion, however, suggests that it should be a proactive process commencing when the client requires the wheelchair on a daily basis, even though they may still have the ability to stand and walk for short distances. The adage "prevention is better than cure" is an appropriate inception in this case.

The use of a wheelchair is generally required when proximal muscle weakness is clearly exhibited in frequent falling, difficulty rising from a seated position and/or an inability to make small postural changes to aid balance when walking over uneven ground or negotiating ramps, slopes and curbs.[8] Although the client may be able to maintain an upright posture against gravity for short periods of time while seated, a kypho-scoliotic asymmetric posture tends to develop as muscles prematurely fatigue when the position is maintained for longer periods. Postural support, such as a contoured back and seat cushion, can help support the individual during postural changes and may also help the user to return to a symmetrical posture.[8]

6.3 PRINCIPLES OF PROMOTING AN APPROPRIATE SITTING POSITION

As sitting is not simply a passive activity, the therapist may work towards ensuring the wheelchair and seating prescription is configured to be as ergonomically sound as possible. It is essential to promote a symmetrical posture and in seating there is a clear role for postural management to enhance musculoskeletal development.[2] During the growth period, it is important to take account of the fact that deformity will develop more readily if the user is exposed to long periods of asymmetric posture as a result of the effect of gravity on weak postural muscles.[9] Common clinical practice is to work towards a level pelvis in the frontal plane, align the trunk with lateral supports so the head is in midline, and support the elbows and forearms on a tray with arm supports.[8]

[7] Mead J; Patient partnership. *Physiotherapy*. 2000; 86(6): 282-284.

[8] Clark J, Michael S, Morrow M. Wheelchair postural support for young people with progressive neuromuscular disorders. *International Journal of Therapy and Rehabilitation.* 2004; 11 (8): 365-373.

[9] Engstrom B. Ergonomics, wheelchairs and positioning. 1193; first edition. Bromma Tryck AB, Sweden.

Some key principles underpin the achievement of an optimal sitting position:

- Maintenance of an appropriate position of the pelvic girdle.
- Effective pressure management.
- Optimum support for the whole body (*See Chapter 8).*
- Promoting activity and participation outcomes.

It should be noted that these principles are guidelines and there may be exceptions to the rule that an experienced practitioner may change to assist a specific client's posture

1. Maintenance of an Appropriate Position of the Pelvic Girdle

Generally, the pelvic girdle is best supported in the upright, neutral position, without either a posterior tilt that results in 'sacral sitting' or an anterior tilt that adversely increases the lumbar curve.

It should be noted that some of the earlier literature does suggest a degree of anterior tilt may be helpful in locking the facet joints to prevent lateral curvature, however this approach cannot be substantiated with a sound evidence base.[10] In some cases, a very small amount of posterior pelvic tilt can be more comfortable if maintenance of a neutral pelvis is difficult for the user to achieve or maintain for long periods of time.

2. Comfort and Pressure Redistribution

It may be beneficial to carefully consider comfort and pressure redistribution during a wheelchair and seating assessment. The ischial tuberosities, sacral coccygeal area, greater and lesser trochanters and intertrochanteric crests are subject to excessive pressures when the client is in the sitting position.[11]

These high pressures will initially cause discomfort but can also lead to occlusion of the capillaries, which may result in a breakdown of the soft tissue over these bony prominences. Many clients with a neuromuscular condition are unable to change position independently and will therefore rely on technology to assist with pressure redistribution in order to maintain a comfortable and safe posture throughout the day.[12]

There is no doubt that a wheelchair with a tilt-in-space function can substantially help to redistribute pressure. There is uncertainty, however, about the best tilt angle and the length of time the user should stay in a tilted position. These factors are likely to vary from person to person and will depend on the degree of asymmetrical posture, fixed deformity and surgical intervention. It is generally accepted that the greater the degree of tilt, the greater the pressure redistribution under the sacrum and ischial tuberosities and femurs.[13] Frequency of tilt is also important. However, the increase in pressure over the spinous processes and scalpulae during tilt may require monitoring.

Movement between the vertebrae is no longer possible for users who have had spinal fusion surgery. Many users report a build-up of pressure in their lumbar region as the day goes by, and tilting back is likely to help alleviate this feeling.

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[10] Meigen l, Kiyoshi M, Kozo H, Toshiyuki F, Naoichi C. Practical problems and management of seating in Duchenne muscular dystrophy. *Arch Phys Med Rehabil.* 2003; 84: 818-824.

[11] Eagle M. Report on the Muscular Dystrophy Campaign workshop: exercise in neuromuscular diseases. *Neuromuscular Disorders.* 2002; 12 (10): 975-983.

[12] Lacoste M, Weiss-Lambrou R, Allard M, Dansereau J. Powered tilt/recline systems: why and how are they used? *Assistive Technology.* 2003; 15 (1): 58-68.

[13] Dicianno B, Arva J, Lieberman JM, Schmeler MR et al. RESNA position on the application of tilt, recline and elevating legrests for wheelchairs. *Assistive Technology.* 2009; 21 (1): 13-22.

Asymmetrical postures can increase sacral seating pressure and asymmetrical loading of the ischial tuberosities. This can be uncomfortable and also cause pressure ulcers. For users who have had spinal fusion surgery, the loss of physiological lordosis can also contribute to increased pressure under the sacrococcygeum.[14]

It is likely that the most effective way to redistribute pressure is by constantly changing position and readjusting the load, but users with a neuromuscular condition will be unable to do this independently because of muscle weakness. The prescription of wheelchair cushions therefore requires thorough assessment.

Many wheelchair cushions can assist with pressure redistribution. Cushions are only one part of the equation, however, and the tilt-in-space and recline functions on an advanced powered wheelchair can also reduce the pressure under the sacrum and ischial tuberosities.[15] Pressure mapping devices may be helpful when undertaking an assessment.

Deformity is often caused by the effects of gravity on the body where muscle power cannot oppose these forces. Assisting clients with neuromuscular disorders to change position in order to counter these effects is imperative.

[14] Drummond D, Breed AL, Narechania R. Relationship of spine deformity and pelvic obliquity on sitting pressure redistributions and decubitus ulceration. *Journal of Paediatric Orthopaedics.* 1985; 5 (4): 396-402.

[15] Lin W, Pierce A, Skalsky A, McDonald C. Mobility-assisted technology in progressive neuromuscular disease. *Phys Med Rehabil Cin N Am.* 2012; 23; 885-894.

Chapter 7 | Assessment Guidelines

Introduction


Photo: LifeSteps Therapy for Kids

Muscle weakness usually commences in the proximal limb muscles and core muscles of the trunk and pelvis; muscles which are key to postural control. Thus, it is advisable to consider supportive seating as soon as the client requires a wheelchair, even if the wheelchair is initially only required for short distances.

There are no physical markers to accurately predict how quickly a condition will progress; therefore it is prudent to work in a proactive manner. Postural support enables effective use of the upper limbs and maintenance of head control. Even if a client only requires the use of a wheelchair to travel long distances, appropriate seating can help to maintain a symmetrical upright posture and maximize functional abilities.

Many wheelchair dependent clients are extremely sensitive to their seating and are likely to be anxious about change, fearing a loss of independent function. The views of the client need to be considered if any changes are planned. It is usually more acceptable to make small incremental changes to seating over a period of time, than many changes all within the same appointment. If a client is currently able to manage a task, even a small change in position can alter their ability to perform the movement required for the task (for example playing a games console, or controlling a joystick).

It is important to do a comprehensive assessment for each client in order to fully address their specific needs.[1] Functional needs and issues will determine how the wheelchair is set up and what seating components are chosen. Consideration should be given to the client's functional abilities in every environment that they may encounter throughout the day.

7.1 ASSESSMENT PROCESS

1 The **first step** in the assessment process involves information gathering and a discussion of the fit and function of current equipment.

2 The **second step i**n the assessment process is the physical assessment. Physical assessment includes a supine mat assessment during which the following are determined:

- Range of motion of the lower extremities
- Presence or absence of orthopedic abnormalities, fixed or flexible
- Presence or absence of postural abnormalities, fixed or flexible
- Body measurements
- Respiratory function
- Skin health

The supine mat assessment is followed by a seated mat assessment.

[1] Wright C, Casey J, Porter-Armstrong A. Establishing best practice in seating assessment for children with physical disabilities using qualitative methodologies. *Disability and Rehabilitation: Assistive Technology.* 2010; 5(1): 34-47.

3 The **third step** involves a functional assessment.

4 The **fourth step** in the assessment process concludes with a discussion of the goals for the intervention.

Observation of respiratory function is ongoing throughout all phases of the assessment

7.2 INFORMATION GATHERING

The gathering of information is an integral part of the full comprehensive seating assessment. It is important to have a thorough understanding of the client and their needs in order to provide the most appropriate seating system. The following topic areas should be addressed and are best obtained through an interview with the client and/or caregivers. Whenever possible members of the community team should be included in the assessment process. Additional information can be obtained by reviewing the chart or professional reports. It is important to collect this information in order to get the complete picture of the goals of the seating intervention from the client and the full team.

Click here to access Positioning Mobility Assessment worksheet

Background information

- Name.
- Date of birth.
- Address.
- Telephone number.
- Referral source.
- Reason for referral.
- Team members.
- Funding information.

Health information

- Doctor(s).
- Diagnosis (is the condition stable or changing?).
- Previous or upcoming surgeries.
- Medications.
- Orthotics.
- Respiratory issues.
- Nutritional/feeding issues.
- Any history of skin breakdown or pressure sores.

Functional Abilities

- Sensory/perceptual skills: Vision, sensation, pain, visual-perception.
- Motor development: Gross motor, fine motor.
- Method of mobility: Level of independence, method, use of adaptive equipment.
- Transfer skills: Method, ease of transfer, level of independence, #/day, use of adaptive equipment.
- Time in wheelchair: How long does client spend in wheelchair during the day?
- Self-Care skills: Level of independence in dressing, eating, toileting, grooming; use of adaptive equipment.
- Assistive technology: Computer access, environmental controls, need for integration through mobility equipment.
- Cognition: Level of independence, safety awareness, judgment.
- Social/Behavioural skills: Safety, behaviors that need consideration.
- Leisure interests: Client's interests, appropriateness of equipment, issues accessing activities.

Environment

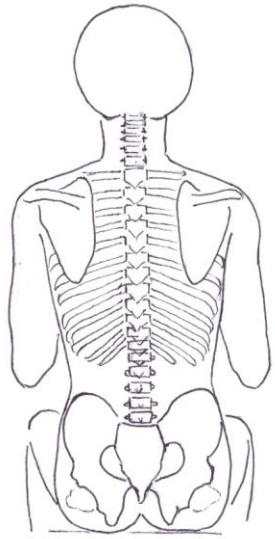
- Daily routine: Consider accessibility issues at home, school, workplace, community e.g., access to buildings, doorways, stairs, bathrooms, tables, desk, computer etc.
- Transportation: Method of transportation, access to vehicle (lift, ramp), type of tie-downs, type of seat belt system, use of public transit etc.
- Seasonal considerations: Extremes in temperature, snow and rain.
- Storage considerations: Ability to charge power wheelchair.

Example: A young man was prescribed a 20" wide wheelchair to accommodate his size. Prior to this the home/school was assessed to ensure that the doorways and hallways were wide enough. Everything checked out. After the final fitting of his chair he went to get on the lift of his van… his chair was too wide!!!

Observation and Physical Assessment in Current Seating System:

- Describe current wheelchair and seating components (including age, condition, and repair history).
- Describe functional positions assumed in the current system to perform functional tasks.
- Discuss and document the client's perception of what is working well and what could be improved.

7.3 PHYSICAL ASSESSMENT



NEUTRAL SITTING POSITION

The 'neutral' sitting position is the ideal goal for positioning in a seating system. This is not always achievable but sitting posture is described in relation to this ideal. The pelvis is the building block of sitting posture. The rest of the body responds to how the pelvis is positioned.

- Equal weight bearing on ischial tuberosities.
- Level ASISs.
- Anterior and Posterior iliac spines level.
- Upright trunk.
- Head in alignment with trunk.
- Hips, knees and feet are at approximately 90°.

Supine Mat Assessment

Range of motion

For a seating assessment we are interested in the client's PASSIVE range of motion at the joints that directly affect the seated position. Thus, all range of motion is done in relation to a NEUTRAL PELVIC POSITION. The pelvis is the base of stability. By measuring range of motion and how it affects the pelvic position we are able to position the client to achieve comfort, stability and function.

Measurements are taken with the client at 90° hip flexion and 90° knee flexion to simulate the "seated position." For example, it is not important to know "true hip abduction" but "hip abduction" as it affects the individual's seated position.

When measuring ROM at the hip:

- It is useful to have a second therapist to maintain the pelvis in a neutral position and note the end of range.
- If a second therapist is not available, it is important to monitor the pelvic position as the hip joint is moved through its range.
- It is important that the hip range not include any pelvic movement.

Click here to access Range of Motion chart

Body Measurements

Measurements are used:

- For sizing of wheelchair.
- For sizing of seating system and components.
- To monitor growth.
- To note any asymmetries.

Measurements are taken:

- Using a tape measure.
- With client on a firm surface.
- First with the client in supine.
- Then with the client in sitting with feet supported.
- On both right and left sides.

Click here to access Body Measurement chart

Seated Mat Assessment

Level of Sitting Scale (LSS) is a global index of sitting ability

LSS is a modification of the Level of Sitting Ability Scale.[2]

- LSS consists of eight levels that are based on the amount of support required to maintain the sitting position and for those who can sit independently without support, the stability while sitting.
- The LSS is similar in concept to the Gross Motor Function Classification Scale (GMFCS). It is a standardized tool to describe/classify a child's sitting ability.
- To assess the level of sitting, the individual is first asked/assisted to assume the sitting position on a high mat or bench with thighs supported to the back of the knees, feet unsupported.

Click here to access Level of Sitting Scale

[2] Green E, Nelham R. Development of sitting ability, assessment of children with a motor handicap and prescription of appropriate seating systems. *Prosthetics and Orthotics International.* 1991; 15: 203-216.

Postural Abnormalities

Individuals with neuromuscular conditions frequently have postural abnormalities. These postures may be fixed (immobile) or flexible. When prescribing a seating system always accommodate for a fixed abnormality and where possible, correct for a flexible abnormality.

Abnormal posture affects the individual's function, may increase the risk of developing a deformity, may compromise function of internal organs and may lead to skin breakdown.

Accurate identification of these abnormal postures is an essential component of a seating assessment. This assessment information is used to determine the individual's postural support requirements in the seating system.

It is essential to do a physical assessment of the individual in the unsupported seated position to determine the effect of gravity on posture. This evaluation should be performed on a firm surface and frequently requires two people to support the individual. This is a very "Hands on" process.

- Observe the posture that the individual adopts on their own
- Determine if the individual can sit hands free or is dependent on external support. This is determined by using the Level of Sitting Scale.
- Observe how limitations of Range of Motion impact the seated position.
- Palpate the individual's bony landmarks. It helps to mark the landmarks with removable stickers. This allows you to visualize postural asymmetries more accurately.
- The sitting posture of the client should be observed from the front, side and back.

Determine if a neutral pelvic position is possible. Begin by placing your hands on the pelvis and move the pelvis to see if a neutral position is possible. Then palpate the ASIS to determine if there is rotation forward on one side. Move the pelvis forward and backwards. Is there a posterior or anterior tilt of the pelvis? Determine if the iliac crests are level or if there is a pelvic obliquity present.

Look at the rest of the body with respect to the neutral pelvis. Observe the body above and below the pelvis. Evaluate the alignment of the hips and lower limbs and then check the trunk, shoulders, head and neck positions. Note any abnormalities *(See Chapter 8 for detailed analysis).*

Observe the spinal alignment of the client in relation to the pelvis. Note any curvatures or rotation of the spine. Determine if the abnormal postures observed are fixed or flexible. If the posture is fixed (i.e., spinal instrumentation) then it will need to be accommodated.

If the posture is flexible:

- How much of the posture is correctable?
- How much pressure is needed to correct the posture?
- Where is the pressure needed to correct the posture?
- Can the client tolerate the amount of pressure needed to correct the posture?
- How does the force required to correct the spinal alignment affect respiration?
- How are functional abilities affected by changes in posture?

Use your hands to determine if a posture is correctible and how much and where the force is needed to correct the posture. Use your hands to simulate where and how much support is needed to comfortably support the individual in a functional sitting position. This information can then be translated into an appropriate postural control system.

The client's position in space will often have a profound effect on their postural control and functional abilities. For example, clients may be able to lean forward to perform functional activities if they are able to tilt their wheelchair slightly to assist in maintaining their balance. Likewise, clients may have improved head control if they are able to lean forward but may not be able to move their head against gravity if they are even slightly tilted. Clients may fatigue during the day so postural supports and use of tilt-in space may change depending on the client's energy level.

Assessment of respiratory function: Assess resting respiratory pattern noting rib cage expansion, diaphragmatic movements, respiratory rate and heart rate.[3] Compare these parameters in lying and sitting. If respiratory function is compromised one may wish to assess oxygen saturation in different positions using a pulse oximeter.

Assessment of pressure distribution: Clients with a neuromuscular condition may not be able to independently change position and may have poor soft tissue coverage of bony prominences. Use of a pressure mapping system on the seat surface or over other areas of point pressure may assist in determining the appropriate postural controls and support surfaces.

Assessment of Function: Take note of the client's level of independence in all functional activities and the use of trick movements or postures used to accomplish the functional activity. For example, some clients with neuromuscular conditions are able to continue feeding themselves after they have lost the ability to lift their hands against gravity by assuming a posture of anterior pelvic tilt and forward trunk flexion to bring their head down to their hands. If the client is using a urinal, there will need to be sufficient room between the thighs for the urinal to be placed.

Functional areas that need to be observed include:

- Eating/drinking.
- Transferring.
- Driving/propelling over a variety of terrains.
- Accessing assistive technology.
- Dressing.
- Bowel and Bladder management.
- Transportation.

[3] Massery M. Breathing and Upright Posture: Simultaneous Needs. *26th International Seating Symposium.* March 11-13, 2010: 25-28.

7.4 GOAL SETTING

Goal setting needs to be done in full collaboration with the client and family. By integrating all assessment information and by understanding the functional needs of the individual appropriate goals for the seating intervention can be determined.

See Chapter 8 for examples of seating and equipment solutions to address postural abnormalities.

Click here to access Positioning Mobility Assessment worksheet

Chapter 8 | Seating and Posture Issues: Possible Solutions

Introduction

This chapter provides additional information for clinicians involved in the provision of seating and mobility equipment. It should be read in conjunction with Chapters 6 and 7 which detail the clinical reasoning for the solutions suggested here.

A full seating assessment is required to determine each client's posture issues or specific difficulties with their current seating systems. The functional implications of supportive seating changes need to be considered for each individual situation.

Modifications to posture and position are best done in small incremental changes as these allow the user to physiologically and functionally adapt to them.

8.1 EQUIPMENT FEATURES AND THEIR EFFECT ON POSTURE

Adaptive seating can maintain symmetrical posture but it can also cause deformity if incorrectly fitted.[1] Posture could be adversely affected if the user is not correctly seated within the wheelchair.[2] Consider the following: components of the wheelchair:

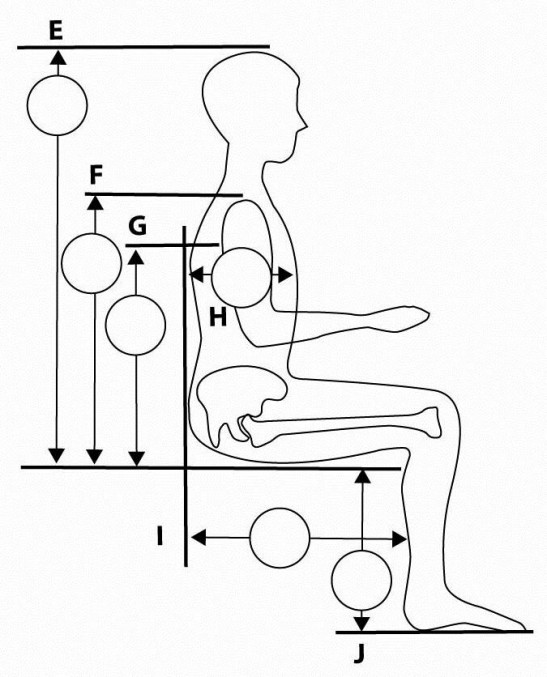


Diagram courtesy of Convaid, Torrance, CA.

- Seat width
- Seat depth
- Seat surface
- Seat height
- Leg/foot support
- Arm support
- Back support
- Head rest
- Wheel position on a manual wheelchair for self-propelling users
- Position and type of joystick

[1] Engstrom B. Ergonomics, wheelchairs and positioning. 1993. First edition. Bromma Tryck AB, Sweden.

[2] Clark J, Michael S, Morrow M. Wheelchair postural support for young people with progressive neuromuscular disorders. *Int J Ther Rehabil*. 2004; 11: 365-371.

Seat Width

Seat width is determined by the **width of the client + 1".** A correct seat width is essential to provide support and comfort as well as to facilitate an optimum position for pushing. An ideal seat width should incorporate the following considerations:

- Wide enough for comfort but supportive enough to ensure pelvic stability.
- Allows arm supports to be close enough to the client to avoid leaning for support.
- Incorrect seat width can contribute to asymmetrical postures.
- Affects positioning of wheels for self-propelling.
- Use of thigh or hip guides may affect seat width.
- Cushion should be matched to seat width.

Seat Depth

Seat depth is determined by the measurement of the **client's thigh length minus ½"-1"**. The seat depth is crucial to ensure that the spine is adequately supported while at the same time maintaining the pelvis in an appropriate position. It is usually best if the seat depth is as long as possible and incorporates the following points:

- Long enough to support the full length of the thigh with the pelvis in a neutral position.
- Provides the maximum sitting area for support, pressure redistribution and stability without impacting on the back of the leg.
- Allows the client to have optimal contact with the back support.
- Seat depth may be influenced by: femur length, pelvic orientation, hamstring contractures, use of orthoses, pressure distribution, leg rest hanger angles, and type of transfer.
- Growth potential should be built into the seat depth.

Seat Surface

The seat surface is important to provide a point of stability and support as a foundation of comfort and control.

- The appropriate seat surface will support the weight-bearing part of the pelvis (the ischial tuberosities) when the client is sitting.
- The seat surface should contour to the pelvis to provide pressure redistribution, support and comfort.
- The weight of the cushion needs to be taken into consideration if the client is using a manual wheelchair or transferring the cushion.
- The weight limit of the cushion also affects the suitability and lifespan of the cushion for the client.
- Consider the immersion properties of the cushion as clients who have very bony prominences can sometimes displace the material and may bottom out.
- Sling seat upholstery can contribute to the development of a pelvic obliquity and is not typically recommended.
- If sling seat upholstery is in place, it needs to be correctly tensioned or the use of a rigidizer or solid seat insert should be considered to prevent a hammock effect.

Seat to Floor Height

The seat to floor height is often determined through consideration of transfer technique and environmental factors such as work surface heights.

- Consider the client's current and future transfer method when determining overall seat to floor height.
- Seat to floor height also impacts the height of the leg rests/footplates and the ground clearance.
- Seat to floor height has an environmental impact as it may affect desk heights, tables, clearance under sinks, entrance into vehicles, etc. if it is too low or too high.
- Seat to floor height of manual wheelchairs may be altered through modification of the axle plate, different wheel sizes, or different cushion heights.
- Seat to floor height on power wheelchairs may be adjusted through the seat frame, use of a different tilt system, different cushion heights, or through the use of a power seat elevator.

Leg/Foot Supports

Adequate leg and foot support is essential to help maintain pelvic stability. The angle of leg and foot support is determined by the comfort range of the knees and feet. Leg rest angle and footplate angle should respect any joint range limitations that a client may have. The seating system should not be used to place a joint in a stretched position. Most people with neuromuscular conditions prefer to sit upright in their wheelchair as this enables them to use their limited ability to reach forward and to maintain head control. To provide stability and maintain position, it may be useful to maintain knee join position close to 90 degrees wherever possible. The neuromuscular population is at risk for the development of flexion and abduction contractures of the hips and knees as well as plantar flexion and inversion deformities of the feet.[3] The following factors should be considered when selecting leg and foot supports:

- Use of angle adjustable footplates may be required if there are restrictions in ankle range or foot positioning.
- Caster interference may occur with the use of tight leg rest hanger angles. 5 cm of ground clearance is recommended so a higher seat to floor height, smaller casters or taller caster forks may be required when used in combination with tight largest hanger angles.
- The height and angle of foot supports require careful monitoring on a regular basis, especially for children as they grow.
- Ankle contractures tend to persist or reoccur so frequent adjustment of foot supports may be required.
- Largest height may affect thigh and foot loading which is an essential part of the seated position. Too little or too much loading can affect circulation and pressure.
- Footrest hanger angle and footrest angle affect the overall footprint of the wheelchair and consideration of the environmental impact (turning circle and transport issues) may be required.
- Consider durability issues when prescribing appropriate foot supports as footplates can be vulnerable and often take knocks from doorframes and other objects.
- If future adjustment is anticipated, providing a multi-adjustable footplate from the start may be cost effective.

[3] Liu, M., Mindeo, K., Hanayama, K., Fujiwara, T., & Chino, N. Practical problems and management of seating through the clinical stages of Duchenne's Muscular Dystrophy. *Arch Phys Med Rehabil*, 2003; 84, 818-824

Arm Support

Arm support height is determined by subtracting 1" from the measured distance between the client's bent elbow and the seat pan. Always measure for arm support height with the cushion in place to take into account any cushion compression.

Photo: Invacare Canada

- The client's arms should be supported by the arm rests so that the shoulders are level and not elevated and the elbows are in a comfortable position.
- Most wheelchairs have adjustable height arm supports. Arm supports with infinite adjustment are easier to adjust to fit than those with adjustments in one inch increments.
- The length of the arm supports can affect client transfers; non-standard or custom fabricated arm pads may be required.
- Full and partial trays can also be used to support the arms, particularly if the user likes to lean forward.
- If subluxation of the shoulder has occurred due to muscle weakness, the arm support may need to be slightly higher than standard to provide more support for both the shoulder joint and shoulder girdle.
- Consider using swivel or mobile arm supports to support functional use of the arms or for activities such as driving.
- Ensure that the trunk is being supported by appropriate lateral trunk supports and not by excessive forces through the arm rests or tray as this not only increases stress on the user's upper limbs and shoulder girdle, but could also encourage undue forces through parts of the wheelchair not designed to withstand them.

Back Support

The back support height is decided by the measurement of the client and level of balance and trunk strength.

- Fit the backrest support for the user with their seat cushion in place.
- Be sure to assess the range and curvatures of the spine and their relation to the pelvis and hips but be aware that maximum backrest contact may not be achievable as this may restrict functional compensatory movement patterns.
- The angle of the back support is critical for ensuring optimal balance, head control, and maximum arm function.
- If the client has a flexible kyphotic posture, the use of some sacral or posterior pelvic support and use of seat angle may to encourage spinal extension can be implemented.
- For users who self-propel and/or can functionally use their arms, the height of the backrest and the amount of lateral support requires careful assessment. Be sure that the back support and the back canes or push handles do not interfere with function or push stroke. For those clients who are self-propelling, the height of the backrest should ideally be below the scapulae.
- Power wheelchair users benefit from support to the level of the shoulders in combination with an adequate head rest support
- Backrests are not generally used to provide head and neck support
- Contoured back supports may not be suitable for users who utilize a powered recline function within their wheelchair because of the alterations to position that take place when the back support reclines and the seat/back angle increases.

Head Rest

A headrest is required for clients who have poor head control or for clients who have tilt and / or recline functions on their wheelchair. The headrest position is dictated by the pelvic and back position and may be impacted by visual restrictions.

- Required for any client who travels in a vehicle while sitting in their wheelchair to protect against neck and whiplash injuries.
- The functional position of the head support may be different to that required for transportation.
- Care should be taken in setting up a head support to ensure the optimum position is achieved.
- Head supports that support under the jaw line are available, but can restrict speech, eating and jaw movement. They may increase risk for aspiration.
- Head support positioning is particularly important with clients who have swallowing difficulties.

Wheel Position on a Manual Wheelchair for Self-Propelling Users

The wheel position on a manual wheelchair affects the stability and maneuverability of the wheelchair.

- The position and combination of wheel size tire type and hand rim on a self-propelling wheelchair requires careful consideration. Undue movement requires an excess of energy expenditure and can increase the risk of repetitive strain injury.
- Larger wheels may be more ergonomically effective.
- Wheelchairs with adjustable wheel positions offer flexibility as the user's needs and posture change.
- Decreased rolling resistance and increased propulsion efficiency can be achieved with a more forward rear axle position.[4] The rear axle should be adjusted as forward as possible without compromising the stability of the user.
- The height of the rear axle position should also be adjusted to improve propulsion biomechanics. The ideal axle height can be measured in two ways.
    1. The angle between the client's upper arm and their forearm should be between 100 and 120 degrees when the hand is resting on the top of the push rim.[5]
    2. While seated in their wheelchair, with their arms hanging down the side, the client's fingertips should be at the same level as the axle.

Position and Type of Joystick

In general, the best position for the joystick is in line with the armrest directly below the client's hand. Many types of joystick are available and all should be thoroughly explored, particularly if hand strength and endurance are affected. Positioning of the joystick can have a profound effect on seating posture. Users will adopt asymmetrical postures and utilize compensatory strategies and movements in order to maintain driving independence. Careful monitoring of drive method is recommended.

- Some manufacturers advocate a centrally mounted joystick to ensure symmetry. These can be mounted on a tray or with a flexible arm but can cause difficulty fitting under tables, during transfers, and for transit.
- The joystick electronics settings can be adjusted to allow increased control and drive ability. Settings such as sensitivity and joystick throw are commonly adjusted with this population.

[4] Brubaker CE. Wheelchair Prescription: an analysis of factors that affect mobility and performance. *J Rehabil Res Dev.* 1986; 23(4): 19-26.

[5] Boninger ML, Baldwin M, Cooper RA, Koontz A, Chan L. Manual wheelchair pushrim biomechanics and axle position. *Arch Phys Med Rehabil.* 2000; 81(5: 608-13.

- According to Pellegrini et al [6] adults with Duchenne muscular dystrophy gradually lose their ability to drive using a conventional joystick but can use mini-joysticks, isometric mini-joysticks, finger joysticks or pads. This study also suggests the individual can use the chin, mouth, or alternative fingers, to control the wheelchair.
- If a user has upper limb weakness and uses a joystick to tilt or recline their wheelchair, their arm can slip back and they may not have sufficient muscle power to move against gravity to re-access the joystick to return to the upright position. An elbow cup or a head switch may be helpful in this situation.

Wheelchair and seating assessments for any individual can be complex and this is felt to be particularly true in conditions where progression is inevitable. Muscle weakness and fixed deformity may represent difficult challenges to both the assessor and the user and collaboration is a necessary element in any assessment.

For children, this is further complicated by growth potential. The following notes are meant to highlight potential seating and posture issues as well as provide possible solutions.

8.2 POSTURAL ISSUES AND SOLUTIONS

Click here to access Postural Problems and Solutions chart

PELVIS

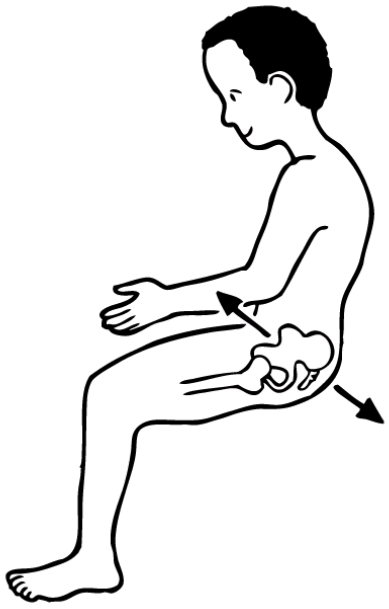
The pelvis is the key point of control in a seated position. Its position affects the rest of the body. Regularly check the position of the pelvis so that any changes can be quickly detected.

Desired Outcomes:

- Stability.
- Facilitate neutral pelvic position while allowing movement as required for function.
- Equal weight bearing on ischial tuberosities.

[6] Pellegrini, N, Guillon B, Prigent H, et al. Optimization of power wheelchair control for patients with severe Duchenne Muscular Dystrophy. *Neuromusc Dis*. 2004; 14(5): 297-300.

Postural Issue: POSTERIOR PELVIC TILT (PPT)

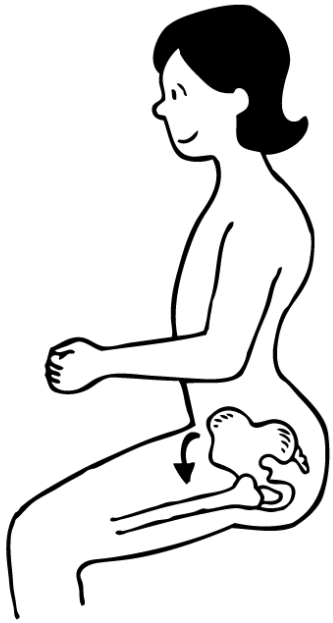


Top of the pelvis is tipped backwards (Posterior Superior Iliac Spine (PSIS) are lower than Anterior Superior Iliac Spine (ASIS).

What you might see: *Posterior pelvic tilt causes the individual to assume a rounded kyphotic trunk posture. The individual may drop his head into forward flexion or will assume a 'goose neck" position to be able to increase his visual field. It is tiring to hold your arms up to do anything functional in this position.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the PPT position is fixed or correctable*
• Physical. • Low abdominal/trunk strength. • Tight hamstrings. • Limited ROM, especially hip flexion. • Fixed pelvic deformity. • Equipment. • Seat cushion too long causing client to slide forward on seat. • Hammocked seat base. • Leg rest/footplates too far forward. • Client not positioned far enough back in the seat. • Pelvic strap positioned too high above ASIS or is too loose. • Incorrect seat to back angle. • Lateral thigh supports limiting abduction thereby restricting anterior movement of the pelvis.	• Use an appropriately angled four-point seat belt to stabilize anterior movement of pelvis. Position pelvic belt to provide firm contact with ASIS. • Consider use of tilt in space or upholstery that provides increased friction to minimize sliding. • Provide cushion or seat rigidizer if using upholstery seat base. • Match leg rest angle to knee range limitations and backrest angle to hip range limitations. • Provide a cushion undercut to accommodate hamstring tightness. • Review transfer techniques and ideal seated position to maximize use of seat depth. • Provide posterior pelvic block/support to limit posterior tilt. • Consider using custom seating to accommodate pelvic position. • Shorten depth of cushion/seat base if cushion length is too long. • Explore alternate cushion materials (foam vs. gel or air) to allow better immersion and stability. • Add contour in seat cushion to provide an ischial block and thigh control. • Position lateral supports to allow hip abduction and promote upright pelvic position.

Postural Issue: ANTERIOR PELVIC TILT (APT)

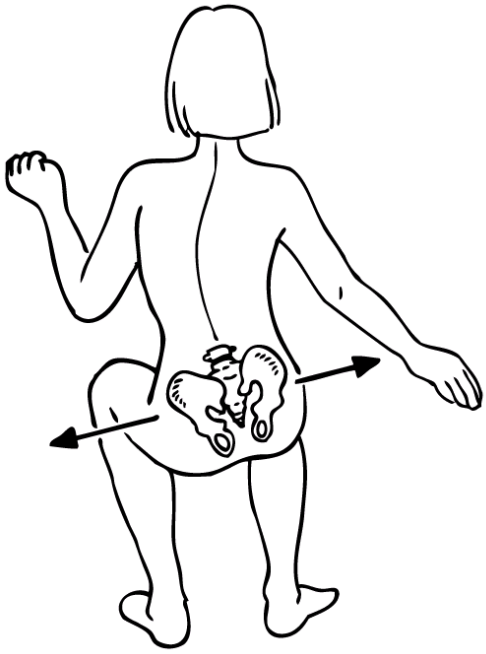


Top of the pelvis is tipped forwards (Anterior Superior Iliac Spine (ASIS) is lower than Posterior Superior Iliac Spine (PSIS)).

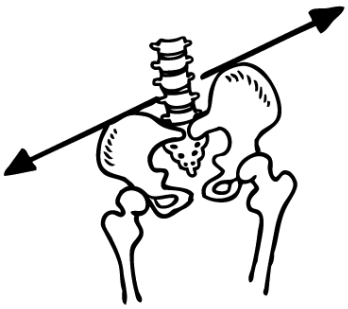
What you might see: *Anterior pelvic tilt causes increase in lumbar lordosis.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Low abdominal/trunk strength. • Tight hip flexors or abdominals. • Lordosis. • User may find this position provides stability and a functional advantage. **Equipment** • Backrest angle may be too tight. • Orientation in space not appropriate for functional use (may limit ability to use compensatory movements). • Anterior tilt of seat base or cushion.	• Use of pelvic positioning belt across ASIS. • Anterior trunk support (corset/wide belt or Tray with custom support). • Chest straps, shoulder straps or abdominal binder to provide anterior stability. • Accommodate APT with custom seating or anterior wedge or have rear seat pan to be lower than front (seat drop). • Tilt may be beneficial. • TLSO or bracing. • Open seat to back angle. • Posterior tilt of base.

Postural Issue: PELVIC OBLIQUITY



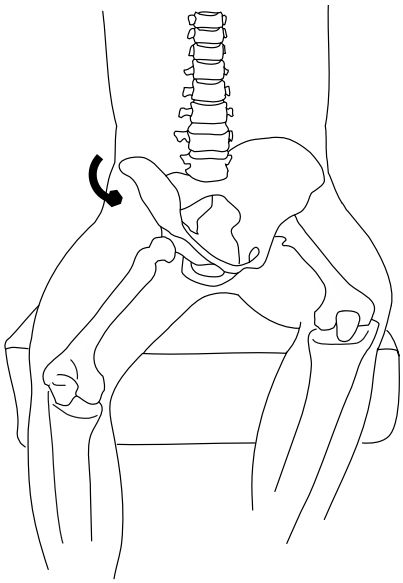
One side of the pelvis is higher than the other (height discrepancy between right and left Anterior Superior Iliac Spine (ASIS)).



What you might see: *Asymmetrical posture and weight bearing with a detrimental effect on sitting balance, respiration and digestion as well as increased risk of tissue breakdown.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Muscle weakness (low abdominal/trunk strength, head and neck weakness, upper extremity weakness). • Scoliosis. • Subluxation/dislocation of hips. • Decreased unilateral hip flexion. **Equipment** • Joystick position/drive method. • Seat too wide. • Seat or backrest too narrow. • Armrests too far apart or too low. • Sling upholstery or worn seating cushion. • Insufficient trunk and head support.	• Adjust seat cushion (add support under low side to correct and under high side to accommodate). This can be accomplished through use of a wedge, different foam densities, using the features of the cushion or with custom seating. • Ensure sufficient lateral trunk support. • Provide pelvic positioning belt. • Consider alternate position for joystick or alternate drive methods. • Ensure correct seat dimensions, arms support positioning, solid seat base and appropriate cushion. • Ensure sufficient lateral trunk support. • Ensure sufficient head support.

Postural Issue: PELVIC ROTATION



One side of the pelvis is more forward than the other (one Anterior Superior Iliac Spine ASIS is posterior/anterior to the other). The rotational deformity of the pelvis originates from the spine. A different degree of rotation may be seen at the shoulder girdle and should also be assessed.

What you might see: *Apparent discrepancy between leg lengths due to pelvic rotation.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • ROM limitations in spine, pelvis, or hips. • Leg length discrepancy or hip dislocation. • Muscular strength imbalance. • Rotoscoliosis. **Equipment** • Failure to accommodate leg length discrepancy or adduction/abduction contractures. • Seat width may be too wide or joystick position may be inappropriate. • Backrest may be too narrow.	• Ensure seat depth matches leg length on both sides. • Provide adequate trunk support. • Provide adequate pelvic positioning belt (lower than the ASIS on forward side). • Ensure seat width matches hip width or provide hip/ thigh guides. • Correct joystick position to provide stability when driving. • Provide cushion rigidizer or solid seat base.

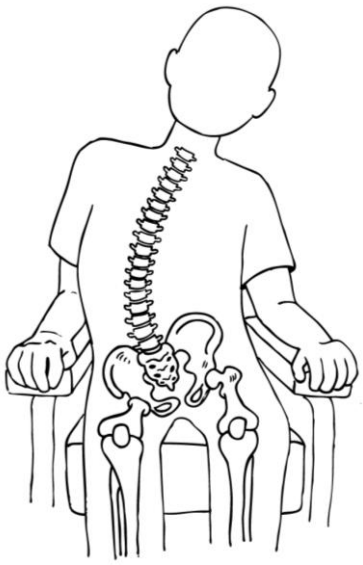
TRUNK

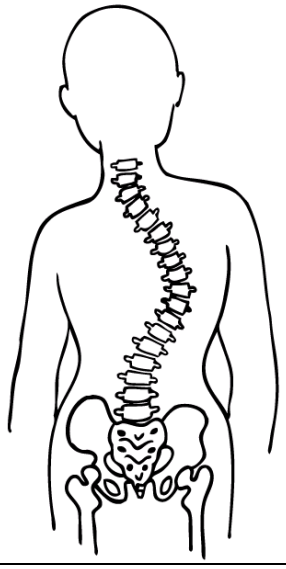
The position of the trunk dictates head position and visual field. Adequate trunk balance is required to free the hands for functional use.

Desired Outcomes:

- Upright trunk.
- Normal lumbar curve.
- Head above shoulders and shoulders above pelvis.
- Functional visual field (eyes level).

Postural Issue: LATERAL TRUNK FLEXION OR SCOLIOSIS





Scoliosis is an abnormal lateral curvature of the spine.

It may be C-shaped or S-shaped and often has a rotational component and may be flexible or fixed.

It is impossible to prevent scoliosis through wheelchair seating alone.

What you might see: *Lateral trunk flexion or scoliosis results in asymmetry of the shoulders and may affect the individual's visual field as the head tilts. May result in asymmetry of the pelvis and consequent asymmetrical weight bearing with an increased risk for tissue breakdown. With severe cases respiration and digestion may be compromised.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Muscular strength imbalance. • Habitual posturing for functional activity or stability. • Mechanical due to spinal rods or muscle contracture. • Pelvic obliquity. • Hip subluxation or dislocation. **Equipment** • Seat width may be too wide or joystick position may be inappropriate. • Hammocked seat base. • Backrest too wide or providing insufficient support.	• Ensure correct armrest height. • Provide adequate trunk support and pelvic support. • Ensure seat width matches hip width or provide hip/thigh guides. • Provide seat rigidizer in cushion or seat base and accommodate or correct for pelvic obliquity if present. • Use three point positioning system – pelvic support, lateral support at apex of convexity and contralateral support above concavity. • Ensure headrest is assisting with midline head positioning. • Consult with Orthotist for consideration of TLSO. • Referral to physician to explore medical or surgical options. • Explore use of lateral tilt. • Consider custom contoured full seating system to provide accommodation and/or correction as required.

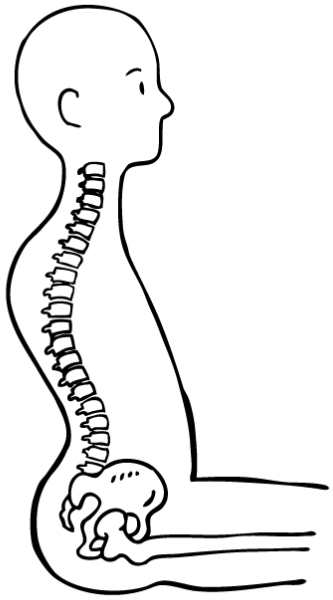
Postural Issue: KYPHOSIS

Kyphosis is excessive forward flexion of the spine.

What you might see: *Limits visual field and head range of movement. May affect swallowing and salvia management in severe cases.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Abdominal or trunk extensor weakness. • Fatigue. • Compensation for posterior pelvic tilt. • Functional seated posture to increase pelvic stability. • Disc degeneration or bone density. **Equipment** • Armrests too low. • Sling back. • Seat depth too long. • Seat to back angle too upright. • Leg rest or footrest too far forward causing posterior pelvic tilt.	• Open seat to back angle. • Ensure adequate seat depth. • Correct posterior pelvic tilt if possible. • Utilize tilt. • Provide adequate trunk support as required. • Provide sacral block to encourage trunk extension. • Provide anterior trunk support (adequate armrest height, tray, chest strap, shoulder strap or curved trunk laterals). • Ensure appropriate position of headrest. • Accommodate through contouring of the backrest/back angle, if fixed. • Match footplate/footplate angle to knee and ankle range.

Postural Issue: TRUNK EXTENSION OR LORDOSIS

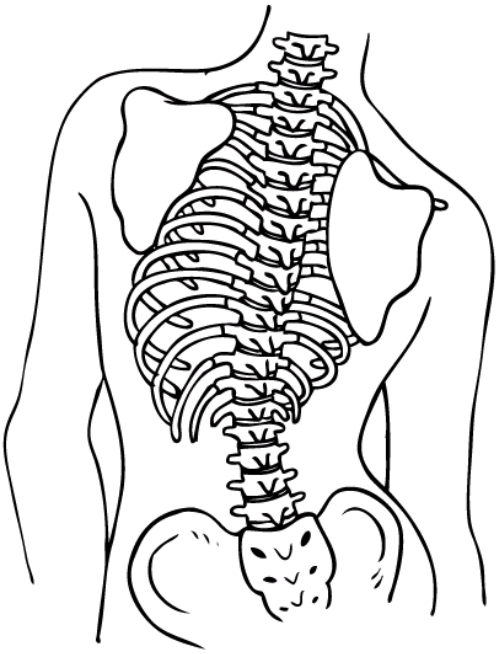


Lordosis is an excessive hypertension of the lumbar spine often combined with anterior tilt. This position is difficult to correct. Use of small incremental changes to posture and position are best as these allow the client to physiologically adapt to them. "Filling in" the lumbar gap is rarely effective and can exacerbate the hyperextension of the lumbar spine.

What you might see: *The individual may retract their shoulders to help stabilize their position in space and maintain an upright posture. The individual may also extend their neck to maintain their balance. If a significant lordosis is present the individual may have to support himself with one arm in order to free up the other arm to reach.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Low abdominal/trunk strength. • Tight hip flexors or quadriceps or abdominals. • Surgical history (mechanical rods). • Fixed spinal deformity. • Functional advantage/balance. • Diaphragm pressure/breathing. • Obesity. • Seeking external bases for support for activities. **Equipment** • Backrest too upright or planar. • Armrests too high. • Excessive lumbar contour. • Footplate too low (knees lower than hips). • Seat depth too long. • Ineffective pelvic control.	• Provide adequate pelvic control. • Provide adequate backrest contouring but do not try to fill in the lumbar space. • Provide back and head support for rest or tilt position. • Consider seat dump or wedge under the cushion. • Consider opening hip angle or use recline to encourage incremental changes. • Consider provision of anterior trunk support. • Provide adequate foot support to have knees level with seat. • Provision of tilt in space to vary the effects of gravity.

Postural Issue: TRUNK ROTATION



Rotation is torsion of the spine around its axis. Trunk rotation is often seen in combination with lateral trunk flexion and pelvic rotation.

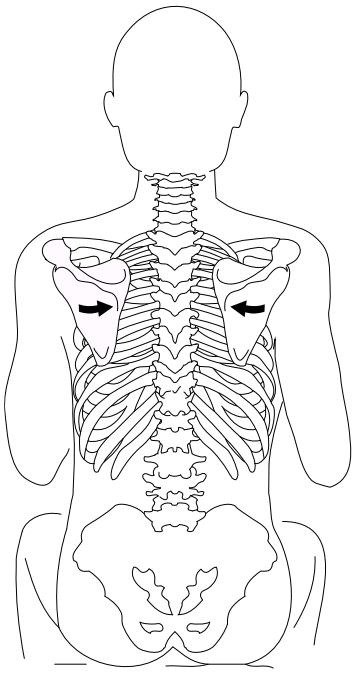
What you might see: *One shoulder will be forward of the other shoulder. The individual may rotate his head in the opposite direction of the rotation in order to maintain a forward eye gaze.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Low abdominal/trunk strength. • Leg length discrepancy or hip dislocation. • Surgical history (mechanical rods). • Fixed spinal deformity. • Functional advantage/balance. • Seeking external bases for support for activities. **Equipment** • Backrest too narrow. • Seat depth too long. • Footplate too low (knees lower than hips). • Ineffective pelvic control. • Leg length discrepancy not accommodated in seating.	• Provide adequate pelvic control. • Provide adequate backrest contouring. • Accommodate leg length discrepancy. • Provide adequate foot support to have knees level with seat. • Use anterior supports on forward side. • Consider placing pelvis asymmetrically in seating system to right trunk and head (for fixed deformities). • Use molded or contoured backrest to accommodate or correct rotation.

Desired Outcomes:

- Upright head.
- Head above shoulders and shoulders above pelvis.
- Functional visual field (eyes level).
- Neutral protraction.

Postural Issue: SHOULDER RETRACTION

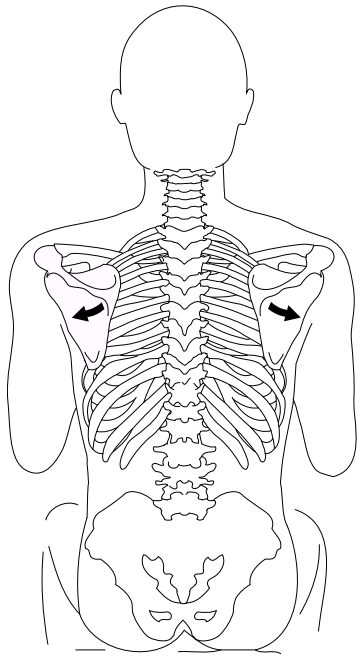


Scapulae are towards the midline or the spine.

What you might see: *The shoulders are pulled into a rear position. Usually causing the chest to protrude and a spinal lordosis to occur. The individual often assumes this position to stabilize his trunk and maintain balance.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Muscle imbalance. • Trunk hypotonia. **Equipment** • Armrests or tray too high. • Backrest or orientation too upright. • Armrest too wide apart.	• Provide adequate backrest contouring with wedges behind scapula. • Consider use of anterior chest support. • Review seating orientation to provide stability. • Provide adequate armrest or tray placement. • Use elbow blocks.

Postural Issue: SHOULDER PROTRACTION



Scapulae are moved away from the midline or spine.

What you might see: *The shoulders appear in a rounded, forward position. Often seen with a kyphotic spine and head in a forward flexed position. Visual field may be affected.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Muscle imbalance. • Trunk and neck hypotonia. • Kyphosis. • Shoulder dislocation. **Equipment** • Armrests or tray too low or too far forward. • Backrest or orientation too upright. • Armrest too wide apart.	• Provide adequate backrest contouring. • Consider use of anterior chest support. • Review seating orientation to provide stability. • Provide adequate armrest or tray placement. • Widen armrests or arm trough.

Desired Outcomes:

- Upright head.
- Normal cervical curve.
- Head above shoulders.
- Functional visual field (eyes level).

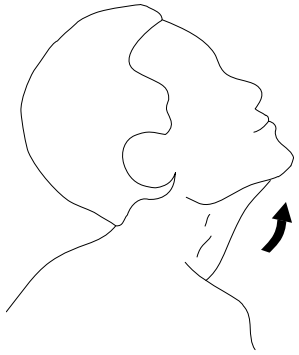
Postural Issue: FORWARD FLEXION

Head tipped down towards chest.

What you might see: *Head is in a forward flexed position with eyes looking downwards. This position is not conducive to social interaction. May see this in combination with a kyphosis.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Muscle imbalance. • Trunk and neck hypotonia. • Kyphosis. **Equipment** • Armrests or tray too low or too far forward. • Backrest or orientation too upright. • Armrest too wide apart. • Headrest too far forward.	• Review seating orientation to provide stability. • Provide adequate armrest or tray placement. • Widen armrests or arm trough. • Consider use of occipital support either in conjunction with headrest or through neck collar. • Provide adequate backrest contouring. • Consider use of anterior chest support.

Postural Issue: HYPEREXTENSION

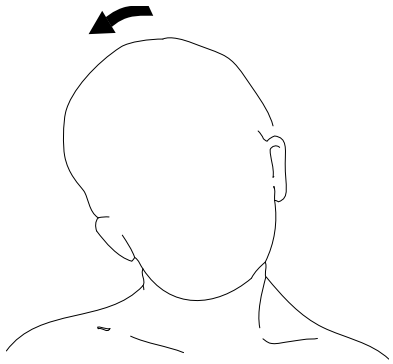


Head tipped backwards with chin poking forward.

What you might see: *Head is tilted backwards with eyes looking upwards. Ultimately could result in reduced neck range of motion.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Muscle imbalance. • Trunk and neck hypotonia. • Lordosis. **Equipment** • Backrest or orientation too upright. • Headrest too far back or too low.	• Provide adequate backrest contouring. • Review seating orientation to provide stability. • Review headrest positioning.

Postural Issue: LATERAL OR SIDE FLEXION



Head tipped to either side with one ear moving towards shoulder.

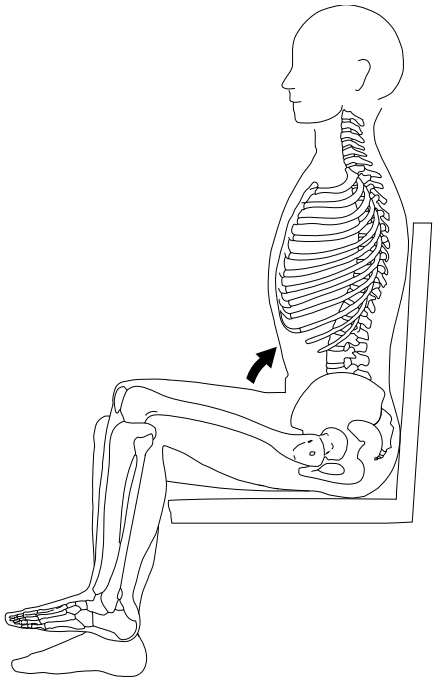
What you might see: *Head is tilted to one side. Visual field will be affected.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Muscle imbalance. • Trunk and neck hypotonia. • Scoliosis. **Equipment** • Headrest too far forward.	• Provide adequate backrest and cushion contouring. • Review seating orientation to provide stability. • Review headrest positioning with lateral or occipital components.

Desired Outcomes:

- Thighs abducted and symmetrical.
- Stability, thighs horizontal and supported.

Postural Issue: HIP FLEXION CONTRACTURES



Flexion contractures at the hips can occur at an early stage, even in users who are ambulatory. Hip flexion contractures often occur with an anterior tilted pelvis. Some clients feel more stable sitting with a closed hip angle (i.e., less than 90 degrees between back and seat).

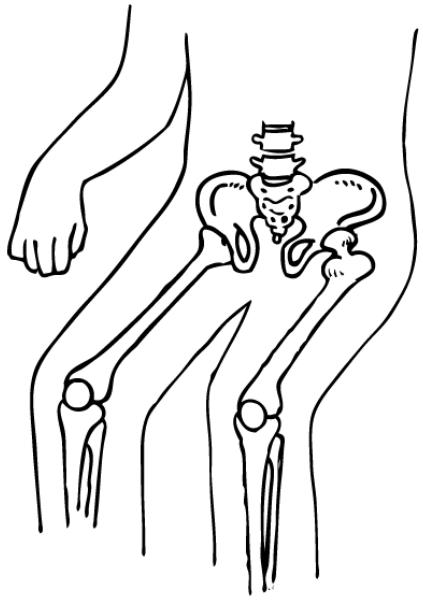
Therapeutic standing and use of the recline function on a wheelchair can help maintain range of movement in the hip. Physiotherapy is also vital.

Consider bone density when prescribing a wheelchair with a standing function or standing frame. Testing of bone density is needed if the user has not been weight bearing for some time or has used/is currently using steroids.

What you might see: *Knees may be higher than the hips in the sitting position. May have spinal lordosis and anterior pelvic tilt. May affect balance for function in sitting.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Hip flexion contracture. • Hip dislocation. • Proximal muscle weakness. • Shortened hamstrings. **Equipment** • Foot plates too high.	• Firm base of support. • Ensure proper footrest height. • Pelvic strap. • May benefit from the use of tilt in space or recline feature on the wheelchair to encourage more trunk extension. This should be used early to avoid excessive hip contractures.

Postural Issue: WINDSWEPT HIP POSITION

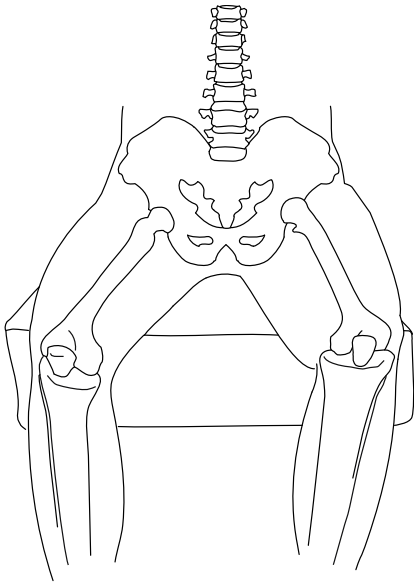


A windswept posture (one leg is abducted and the other is adducted) is often associated with pelvic/spinal rotation.

What you might see: *One leg will be abducted and may be resting against the side of the wheelchair; the other leg will be adducted and frequently internally rotated. The adducted leg may be in contact with the abducted leg.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Pelvic rotation. • Scoliosis. • Dislocation of hip. **Equipment** • Sling seat. • Foot rest position too high. • Leg length discrepancy not accommodated. • Seat too wide or too narrow.	• Firm base of support. • Ensure proper footrest height. • Pelvic strap. • Medial thigh support (pommel). • Lateral thigh support. • Contoured custom cushion to accommodate fixed deformities. • Ensure seat depth matches leg length on both sides. • Ensure seat width matches hip width.

Postural Issue: HIP ABDUCTION



Both hips are abducted and usually externally rotated.

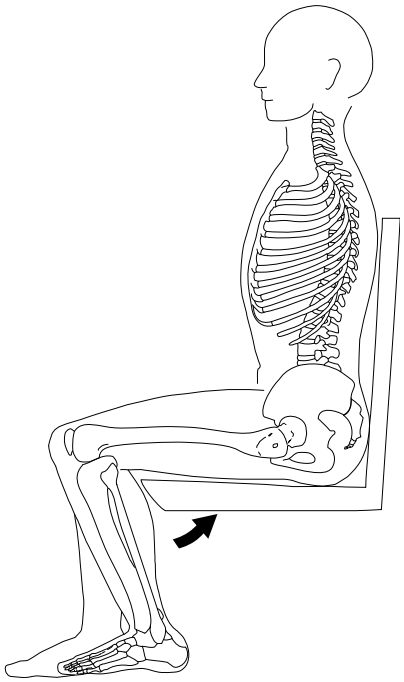
What you might see: *Legs will be abducted and either resting against the sides of the wheelchair/seating system or if there are no sides to the chair the legs may be overflowing the edge of the chair. If the abduction is significant then the individual may have to adduct his lower legs and rest on the sides of his feet to maintain his feet on the footrests.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Low muscle tone. • Weak muscles. • Contracture of the abductor muscles. **Equipment** • Foot rests too high. • No lateral pelvic or thigh supports. • Seat too wide. • Seat depth too short.	• Firm base of support. • Ensure proper footrest height. • Ensure seat width is appropriate. • Provide lateral pelvic and thigh support. The thigh support may have to extend over the edge of the seat to the outer edge of the knee to prevent excessive abduction and maintain comfort. • Accommodate for abduction contracture but ensure adequate support so that the contracture does not increase.

Desired Outcomes:

- 90 degrees of flexion.
- Accommodate when 90 degrees of flexion is not possible.
- Decrease tension in hamstrings that might increase a posterior pelvic position.

Postural Issue: FLEXION CONTRACTURES



Flexion contractures at the knees usually occur once an individual becomes a wheelchair user.

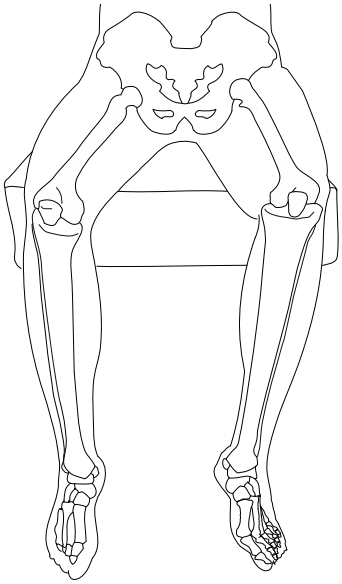
What you might see: *Will often see an individual rest his feet at the back of the footrest on the heel loops or top of the footrest to relieve the stretch on the knee. May also see the individual drop his feet off the side of the footrest for this same purpose.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Muscle weakness. • Shortened hamstrings. • Weak proximal muscles. • Position of function and stability, often combined with forward trunk flexion. **Equipment** • Seat too short. • Footrest too high.	• Ensure proper footrest height. • Select an appropriate hanger angle and footplate position to match the client's range of movement. • Seat depth may need to be shortened and the cushion may need to be undercut. • Consider the pressure exerted by heel and calf straps. • Referral to physician to explore medical or surgical options.

Desired Outcomes:

- Foot supported in a plantar grade position.
- Prevent pressure to feet and to ischial tuberosities.

Postural Issue: PLANTAR FLEXION (POSSIBLY WITH ANKLE INVERSION)



Contractures of the feet often occur once the individual is no longer standing or weight bearing. It is extremely important to promote the use of Ankle Foot Orthosis (AFO) early in the intervention process to help maintain the range of motion of the ankle.

What you might see: *The individual will often rest on the sides of his feet as he cannot rest them flat on the footrest. He may also drop his feet off the sides of his footrest to avoid pressure on the feet.*

Possible Causes or Contributing Factors	Suggestions for Intervention *Management will depend on whether the position is fixed or correctable*
Physical • Dorsiflexion muscle weakness. • Achilles tendon shortening. **Equipment** • Footrest too low or feet unsupported in the wheelchair.	• Ensure proper footrest height. • Angle-adjustable footplates (anterior/posterior and medial/lateral). • Correct size/shape footplates. • Padded foot box may be needed to protect feet. • Ankle huggers or additional foot straps. • Referral to physician to explore medical or surgical options.

Chapter 9 | Features and Benefits

Introduction

This chapter includes information about manual and powered wheelchair product solutions that might help people with neuromuscular conditions.

Medical equipment vendors and representatives of assistive technology companies who provide equipment demonstrations and/or help with provision should be made aware of the clinical issues in the long-term management of neuromuscular conditions. Collaboration between the assistive technology provider and the clinician can produce the best outcome for a client.

> *"The functions on my new powered chair have given me much more independence, freedom and mobility. The sit to stand function has allowed me to stand and maintain my limited walking ability. In addition, this feature means I can also have direct eye contact at any level. For me, as a conference speaker, this is essential."*
>
> **— Michael McGrath,**
> **Inspirational Speaker, Leader and Adventurer**
> **Only person with muscular dystrophy to walk to both the North and South poles**

Mobility Devices and Optional Features

Product Type	Specific Options	Benefits	Drawbacks
Stroller	• Simple umbrella fold. • Tilt-in-space. • Various chassis options.	• Lightweight strollers can be useful for a family with several children or who need space in a vehicle to transport other equipment. They are often good as a secondary piece of equipment to aid family lifestyle. • Often have basic modular seating options (i.e., laterals, lumbar supports) for better seating positions. • Option for forward or rearward facing seats on some models. • Can be very useful for families who are not yet ready to accept a wheelchair. • It can also be used as a transport system (i.e., car seat). The need for fewer pieces of equipment can resolve complex funding issues and may suit a young child who doesn't settle well in different pieces of equipment. • Child can stay in the seating system and be transferred onto a home base, a school base, a wheeled base (i.e., stroller type wheels). • It may be easy to push and often more compact and lighter than a wheelchair.	• Not very supportive, can only add minimal support or harness. Posture is often compromised for lifestyle needs. • This more comprehensive stroller type is not as lightweight for families, and often cannot fold down compactly. Only some models are crash tested. • Families of school age children should be encouraged to investigate wheelchair options to foster a degree of maturity within the child. This can be emotionally challenging for some parents but allowing children to independently propel will improve spatial awareness, decision making and cognitive ability. • The danger with this is that the child is not getting frequent changes of position. If the seating is not ideal than they have not got an alternative to change into, also make life difficult when child has to go into bigger seating etc., they then effectively outgrow everything at once.
Manual Wheelchair	Standard self-propelling.	• Independence. • Essential for longer distances/outside use as walking and exercise tolerance deteriorates. • Ease of transportation in car.	• Fatigue. • Potential for repetitive strain injury (RSI) to shoulders, elbows and/or wrists. • Limited availability of sizes which may require adaptation to match the user's needs. These chairs are mostly made of steel or chrome and are therefore not lightweight.

Product Type	Specific Options	Benefits	Drawbacks
Manual Wheelchair (continued)	Mobility device pushed by caregiver.	• Ease of transportation in car. • Essential as back up when powered chair is main form of mobility.	• No independent mobility. • May not suit caregiver's needs.
High-performance Manual Wheelchair	Many models available with rigid or folding frames, and a range of accessories.	• Custom fit. The choice of seat width, depth and height, back support, arm support, foot support, height and leg support angle ensures that the wheelchair matches the user's size, providing appropriate postural support and discouraging the development of deformities. • Ease of propulsion/control. The adjustability offered by a high performance manual wheelchair, particularly in the positioning of the rear wheels, ensures a good pushing position. The weight distribution between the rear wheels and castors makes this type of wheelchair easier to maneuver and 'back wheel balance' than a 'standard' manual chair. It increases independence for all users, particularly those with upper limb weakness. • Seat height. The seat height can be made higher than in a 'standard' wheelchair, by lowering the position of the rear wheels and modifying the front fork and castor housing. This can greatly benefit users who have difficulty rising from sitting to standing or for environmental access. • Exercise. Self- propulsion ensures a user is undertaking some physical activity even if he or she cannot walk. This benefits general health and assists with weight control. Active use of the arms may help maintain strength. • Increased independence. The ability to self-propel alleviates the need for the user to be accompanied. He or she may be able to dismantle the wheelchair independently and put in a car. • Flexibility. This type of wheelchair can be adapted to meet the changing needs of a person with a progressive condition. It alleviates the need for frequent changes of equipment although the set-up of the chair should be reviewed regularly. It is possible to add items such as power-assist to some models, once the user loses the ability to independently self-propel.	• Initial cost. • The wheels will need to be set up accordingly, allowing for growth and with the ability for adjustment when the current dimensions are outgrown. This may not be as aesthetically pleasing but will be cost effective.
Power Assist Self-propelling Wheels	Power-assist self-propelling wheels.	• User still 'exercising' and using remaining muscle strength. • Manual and power assist wheels to be used on same base can be very versatile and decrease storage /transportation issues. • Limits repetitive strain injury (RSI) issues. • More than one power setting for use in different environments (depending on model). • Can be added to manual wheelchair when required (e.g., end of day or over long distances).	• Limited range muscle strength. • Heavy to take on/off. • Batteries expensive to replace. • Need to check with manufacturer regarding compatibility and warranty. • Initial cost.
Power assist on manual wheelchair	Joystick control.	• Can be added to manual wheelchair when required (e.g., end of day or over long distances). • Easier to transport than powered wheelchair. • Two products in one. • Can be funded by second agency. • Possibly less psychological adjustment required.	• Limited range. • Less stable, especially on ramps. • Heavy to lift in/out of vehicle. • Need to check with manufacturer regarding compatibility and warranty.
Powered Wheelchair	Powered chair controlled by joystick.	• Offers independence over a larger area than possible with manual propulsion. • Allows user to keep up with peers and facilitates social integration. • Less fatigue and repetitive strain injury (RSI). • Powered seating options available on some models e.g., tilt-in-space, seat lift, powered recline and leg supports. • Often allows interface with more supportive seating systems. • Drive profiles are programmed to each individual user at set up. • Some joystick controls can be upgraded to more sophisticated systems. Consideration needs to be given to this at the time of issue/purchase.	• May be difficult to transport in car due to weight. • Asymmetric position of joystick (at end of armrest) might lead to postural asymmetry. • Consideration needs to be given to Health and Safety issues. A risk assessment and driving test may be required. • Maintenance costs higher than manual. • Regular maintenance required. • Insurance advisable.

Product Type	Specific Options	Benefits	Drawbacks
Tilt-in-Space	Powered.	• User can select appropriate posture for different activities. • Can reduce the influence of scoliosis. • Uses gravity to help maintain head and trunk control, reducing need for intrusive head supports. • Enables the user to independently change his or her position. • Can help reduce the development of neck contractures (which cause decreased function, poor posture and discomfort) if used with a suitable supportive headrest. • User can rest against the backrest and avoid a dysfunctional forward leaning posture. • Can reduce the need for intrusive, and often uncomfortable, trunk supports, thoracic pads, harnesses and vests. • Increases pressure redistribution on the backrest and head support, reducing the load on the user's seating area. • Aids tissue viability and reduces the risk of pressure ulcers. • Reduces muscle fatigue and prolonged sitting in one position which can cause postural pain. • Aids management in recovery following spinal surgery. • Improved sitting position does not compress internal organs, which helps respiration, digestion and elimination. • Reduces the need for regular transfers (and moving and handling) to lie down for rest or relaxation, which can interrupt daily life, be socially isolating or impossible to achieve. • Can help caregivers to position user in the back of the seat when transferring into a chair. • Allows for variants of posture to allow function, stability or rest depending on the activity.	• Cost. • Possible incompatibility with user's equipment e.g., mobile arm support. • May increase seat to floor height.
Recline	Powered.	• Recline systems provide a change in orientation by opening the seat-to-back angle and, used with elevating leg rests; also opens the knee angle. • The negative recline function (closing up the hip angle) can be used with tilt-in- space to provide fine adjustments to posture, enabling good head and sitting balance, stability and eye contact. • Can help maintain Range of Movement at hips and reduces risk of hip contractures developing/progressing. • Varying the hip angle throughout the day can reduce any pain and/or stiffness. • Powered recline can assist with toileting in the wheelchair.	• May cause loss of good postural positioning. • The use of tilt-in-space may alleviate this problem. • Cost. • Increases the bulkiness of the backrest pan.
Seat Elevation	Powered.	• Increased access in different environments. • May minimize the need for some adaptations to environments. • May improve eye contact with peers/colleagues. • Helps compensate lack of lifting & reach ability in the arms.	• Cost. • May increase seat height so impacting on the access to the user's environment. • May increase seat to floor height impacting on access to user's vehicle and environmental access under work surfaces.
Elevating Leg Supports	Powered.	• Change of position can improve comfort. • Beneficial for circulation. • Can be used to decrease risk of contractures in knees and hips. • Can change pressure distribution through feet.	• May not provide adequate leg support for users with knee contractures. • May have issues with contact between the user's leg and the hanger components. • Cost. • Require regular maintenance.
Standing Chairs	• Manual. • Powered. • Manual and powered combination.	• Facilitates functional, purposeful standing during activities. • Reduces transfers, and moving and handling issues for caregivers/families. • Standing without using large pieces of equipment (e.g., static standing frame) allows more dignity. • General benefits of standing. • Facilitates natural symmetrical standing posture. • Develops, improves and maintains upper body balance and strength. • Improves/maintains range of movement in spine, hips, knees and ankles.	• May cause loss of good postural positioning. • Possible discomfort. • Excessive force on joints. • Cost.

Product Type	Specific Options	Benefits	Drawbacks
Standing Chairs (continued)		• Provides opportunities for prolonged stretching which helps reduce development of contractures. • Changing position reduces risk of pressure issues • Improves systemic functions (bladder, digestive, respiratory, and circulatory). • Lessens progressive scoliosis and assists skeletal development. • Alleviates discomfort by change of position • Beneficial psychological effect.	
Specialist Control Systems	These include: • Different shaped joysticks. • More sensitive joysticks. • Mini joysticks. • Proportional head control. • Alternative switches. • Attendant drive controls. • Switch drive controls.	• Allows continued independent drive control. • Customized control system to suit user's abilities. • Can be programmed to individual's requirements as needs change. • Allow integrated control of multiple types of assistive technology (w/c, tilt, EADL, cell phone). • Can be used for caregiver to provide assistance if needed.	• More complex system. • User needs to learn new skills. • More technical skill required to assess and setup. • Can be harder for care givers to drive if needed. • Potential incompatibility between multiple assistive devices. • Cost.
IntelliDrive or Track Control Technology	Integrated drive control add-on to power wheelchair which decreases the drive demands of the user.	• Compensates for change in grade or slope of the environment to enable the user to maintain a straight forward path without making drive compensations. • Compensates for uneven terrain and surface vibrations. • Increases stability and evenly distributes the weight over the footprint of the wheelchair. • Increases the maneuverability and helps maintain a straight forward path for switch drive controls.	• Cost. • Increase complexity of wheelchair and electronics.
Mobile Arm Supports	Single or pair of height-adjustable mobile arm support(s), in which the forearm is supported in a trough.	• Device enables frictionless, gravity-eliminated movement at a range of heights. • Gives user a wider reach, increasing the range of tasks that can be performed.	• Attached to the back of the user's powered wheelchair or work space. • May affect total width as well as the possible use of tilt-in-space and recline. • May compromise arm or hand stability for functional tasks.
Aids to Daily Living	Different systems available on the market, some of which may need to be fitted onto the wheelchair.	• Enable user to independently perform tasks such as working audio-visual equipment, managing the telephone, turning lights on/off, drawing curtains around the home. • System for monitoring and allowing access to callers.	• May be expensive. • May be complex to use. • Limited to environment. • Technology may become obsolete rapidly. • Technology compatibility issues. • Technology support required.

Chapter 10 | Planning for Different Environments

Introduction

The introduction of the first wheelchair, whether manual or powered, will inevitably raise concerns for clients and caregivers about access to everyday environments such as home, school, university, workplace and outdoors.

Using a wheelchair will have implications for all these environments as well as affecting private and public transport decisions.

Many clients and caregivers will have planned ahead but others must face the realization that changes are needed.

10.1 HOME ENVIRONMENT

Ideally, home adaptations need to happen at the same time as wheelchair provision. An assessment of the home environment is essential to ensure it is compatible with the user's long-term needs. Successful adaptations provide access into and around the house and yard, and include enough space to accommodate any future wheelchair.

The Canada Mortgage and Housing Corporation (CMHC) publishes Accessible Housing by Design guidelines to help users and professionals plan suitable wheelchair accessible environments. The guidelines give advice on long-term housing needs, space requirements and architectural specifications.

The BC Housing Home Adaptations for Independence (HAFI) program provides financial assistance to help people with disabilities in British Columbia to continue to live in the comfort of their home.

The Family Fund helps families in BC who have children and youth up to 19 years of age with special needs living at home by providing one-time grants to support projects such as home renovation. Assistance is also provided for adults living at home who are classified as having a developmental disability.

In the past, powered wheelchairs were considered less maneuverable than manual chairs, as they required more circulation space. However with technological advances and new products constantly coming onto the market this is no longer always the case. Due to upper limb weakness a user may gain more independence in the home by using a powered wheelchair rather than struggling to self-propel.

10.2 SCHOOL ENVIRONMENT

Careful consideration should be given to the school environment of a student with a neuromuscular condition. The wheelchair should be compatible with the school environment, both indoors and outdoors. The needs of a student will vary as the condition progresses and the level of impairment changes. A student who is mobile at the beginning of his or her school life, for example, may be a full-time wheelchair user later.

Forward planning is essential. The school should review and update an accessibility plan including a fire and evacuation plan. This may also be a requirement of the local school board, although it varies from area to area. It may also need to be documented with the Individualized Education Plan (IEP).

Areas to consider in the accessibility plan:

- Access to school entrance and all classrooms.
- Access to, and use of, lifts and elevators: ensure that the lift has adequate space and is safe to take the weight of wheelchair and user.
- Circulation space in classrooms and corridors.
- Ramped access to emergency exits.
- An emergency evacuation plan.
- The playground should be safe enough for the child to independently play and socialize with friends.
- Access to, and space within, the bathroom.
- Access to dining, recreation and all educational areas.

School staff need to be aware of their responsibilities regarding the health and safety of a wheelchair user and his or her relation to other students. They may benefit from advice and support from the student's therapists. School transport should be considered when assessing wheelchairs that have electric functions and are likely to be heavier, higher and longer than standard powered wheelchairs. Check the maximum weight capacity for a bus lift as well as maximum head clearance.

10.3 UNIVERSITY

Consider the college, university and/or campus facilities. Contact the school's Student Support Services as early as possible to discuss the student's needs. Grants may be available for additional equipment. Provision of service varies from school to school but documentation of the type of disability is required at all schools.

10.4 WORKPLACE

Consider the user's work environment when assessing for a wheelchair. The employer may have an ergonomic advisor involved in assessments. Level of support varies between employers. There are funding programs that may assist in provision of technology to attain or maintain employment.

10.5 TRAVEL/TRANSPORT

Although it is recognised internationally that it is safer to travel in a vehicle seat than in a wheelchair, for many people with a neuromuscular condition this may not be possible. In fact, having access to transit is often essential for community access for many people who use a wheelchair. If a user has to travel in a wheelchair, then ideally that wheelchair should have passed a crash test to a relevant and recognised standard. However, it is possible that the most appropriate chair/seating system combination has not been crash tested. This does not mean that the device is not safe; it just means that in that particular configuration it hasn't been yet been proved to be safe. More important than the wheelchair having been crash tested is that the wheelchair is properly secured in the vehicle and the occupant is secured in the chair. Studies show that most injuries to users when in transit are due to non-collision events such as sharp turns or sudden stops and lack of or improper wheelchair securement and or restraints. Injuries also occur from incidents when entering or exiting vehicles.

See RESNA's Position on Wheelchairs Used as Seats in Motor Vehicles for details.

Weak neck and shoulder muscles are common for people with a neuromuscular condition and may result in poor head control. This means they are at higher risk of whiplash injury. Use of an adjustable headrest, possibly with shaped side supports, will help to minimize this risk. Using a slightly tilted position on a wheelchair when accessing a vehicle by ramp may help with head clearance and assist transfers into the vehicle.

Some users may wish to drive from their wheelchair. In most instances the chair is secured to the vehicle via a sturdy pin projecting below the chair that locks into a clamp bolted into the floor of the van. Remember that this only secures the chair, not the occupant and a seatbelt must also be worn which anchors the occupant to the vehicle. This should preferably be a three-point harness type.

It is important that the wheelchair user take responsibility for ensuring their chair is appropriately secured. If tie down points on the chair are not obvious a picture would make it easier for the driver to secure the chair. Alternatively webbing can be left in place on the chair to serve as an appropriate interface between tie down system and chair.

10.6 OUTDOOR ENVIRONMENT

Wheelchair users who use their chairs on the road are at greater risk than other road users and this is not reflected in current road safety laws.

There is currently no standardized training for users to ensure that the increased mobility provided by their chair is not decreasing their safety or minimum fitness standards, such as eyesight standards or other health issues. The wheelchair and the user must be visible in poor light or darkness. Lights and reflective strips for the wheelchair, and reflective clothing for the user, are essential for safety. (Most funders do not provide these.) Lights come as an optional extra on some powered wheelchairs but can be expensive. Bicycle lights are a cheaper option and can be mounted onto the wheelchair frame without the need for drilling. Reflective strips can also be bought at most cycle shops and be used to customize the wheelchair. It is wise to note that lights will increase the users' ability to visualize his or her environment but they do not necessarily make the user easier to visualize. The lights are usually mounted low on the wheelchair frame so they light up the users' path but may not be in the sightline of vehicle drivers.

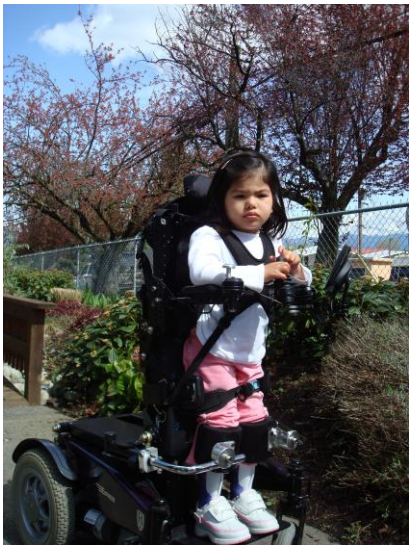
> *"I would really like to have lights on my chair so people can see me when I go out now it's dark, and I would like to paint it red."*
>
> **— Irman, 13 (DMD)**

A person using a wheelchair should follow the rules that apply to pedestrians. For the most part these are covered in the by-laws that were developed by local municipalities rather than the Provincial government. B.C.'s Motor Vehicle Act states that powered wheelchairs do not require licence, registration or insurance. If you are driving a wheelchair you are considered a pedestrian. Although not governed, it is suggested that the user avoid driving a powered wheelchair if he or she is taking any medication, drug or substance that may affect their skills or judgment.

Powered wheelchairs are currently prohibited from using motorways, bike lanes, and bus lanes but may need to use these to access certain environments. You are not allowed on the road if there is a sidewalk available. If there is no sidewalk you should travel on the extreme left hand side of the road facing oncoming traffic.

10.7 SPECIALIZED WHEELCHAIR TRAINING

Life in a wheelchair can be very restricting, especially if the user has to rely on family, friends or caregivers to get around the neighbourhood or even into the yard.



It can feel even more restricting for children as they watch their friends play outside but are unable to join in. All children need to get out and about, play, enjoy the fresh air and spend time with friends. Children who use wheelchairs are no different.

Perhaps more than their peers, they need to move safely around their home and neighbourhood and participate in the daily activities at school and home that others take for granted.

In the street, children who use wheelchairs need to learn how to get up and down curbs, deal with traffic and pedestrians, and negotiate crossings and parked cars. A child needs the confidence and skills to develop his or her individual way of living, and growing, with the wheelchair. They need to learn how to use their wheelchair to reach out and broaden their horizons so they can realize their full potential.

10.8 PUBLIC TRANSPORT

Introduction

Wheelchair users can only take their place, and play a full role, in society if they have good, accessible public transport. The positive effects of accessible public transport cannot be underestimated. Being able to use public transport can improve self-esteem, quality of life, and independence as well as having social and economic benefits.

Accessibility

Public transport has radically changed in recent years and much more is now accessible to wheelchair users. An increasing number of service providers can provide information for users on which services are accessible as well as offering tips on travelling in a wheelchair.

Safe transportation of wheelchair users

Not all types of wheelchair are suitable for public transport. Users with an extra-large wheelchair, stroller or scooter may want to contact the relevant transport operator for help and advice. Some transit services offer access to a training bus to practice access and use of restraints in a non-pressured setting.

Travelling by bus or train

A bus driver may be unable to help a wheelchair user onto the bus but is responsible for securing the wheelchair once inside the bus. There are specified spots for securing wheelchairs on public buses and trains.

Ramps used to board buses and trains can vary in height because of curbs and platforms. Although most wheelchairs have anti-tip devices, care should still be taken when negotiating ramps.

Users should be aware of the limitations of their own wheelchair regarding inclines, sideways tilt and detachable accessories that can impact on safe travel. Users are responsible for ensuring their wheelchair is in serviceable condition, for example tires correctly inflated so that brakes work efficiently and effectively.

Many door-to-door services are also available. Most of these services use wheelchair accessible buses or vehicles that have been adapted by specialist companies. Vehicles should always carry different types of

wheelchair tie-down and occupant restraint systems so that all wheelchairs can be transported safely. When applying user restraints, consideration should be given to the most suitable type, and the correct positioning for the wheelchair user.

Accessories are sometimes needed by a wheelchair user during travel. These should be secured safely and may require padding. Good communication skills are vital to staff involved in transporting wheelchair users, to ensure maximum safety and respect for the user's rights.

Crash testing of wheelchairs in road passenger vehicles

There has been some crash testing of wheelchairs. These involved use of a front impact sled test and dummies to assess the safety of wheelchair users when travelling in road passenger vehicles. These tests have usually simulated an impact at 30 miles per hour. See RESNA's Position on Wheelchairs Used as Seats in Motor Vehicles for details. The dummies used simulated average sized users of the equipment. All the tests show that the most vulnerable parts of the body for wheelchair users are the head and neck. Testers recommended that head and back restraints be fitted in all vehicles carrying wheelchair users. These tests also found that vertical shoulder straps are not suitable for use with belt restraints, as they increase the risk of injury. Lap and diagonal belt restraints are recommended. There are advantages and disadvantages to sitting in a rear or forward facing position, depending on the type and size of vehicle, anchorage variations, restraints, and size of the wheelchair user. Powered wheelchairs cannot always get close to the back and neck restraints, because of battery containers and gears. This gave rise to greater injuries in the tests. The tests concluded that it was impossible for a passenger seated in a wheelchair to be given the same degree of protection and safety as a conventionally seated person. Changes were recommended to vehicles, restraints and wheelchairs but it was accepted that none would be made in the foreseeable future due to costs.

APPENDICES

APPENDIX 1

NEUROMUSCULAR CONDITIONS

The term 'neuromuscular conditions' is used to describe a group of mostly genetic disorders, which are generally progressive. They cause loss of muscle strength and sometimes this deterioration can happen quickly.

There are over 60 types of neuromuscular condition, including muscular dystrophy. The age of onset varies between, and within, conditions. Some conditions, such as Duchenne muscular dystrophy, always begin in childhood while others start to affect individuals in adulthood.

Included here is brief information about the most common neuromuscular conditions. More detailed information is available from the Muscular Dystrophy Canada.

BECKER MUSCULAR DYSTROPHY

Becker muscular dystrophy (BMD) is an X chromosome condition. It affects boys but girls in families are often carriers and may pass the gene onto the next generation. It is a milder variant of Duchenne and those boys/men with the condition experience similar problems to those with Duchenne. The condition varies in severity. It can be almost as severe as Duchenne or mild enough to only be diagnosed later in adult life. Some of those with Becker muscular dystrophy will lose the ability to walk in early adult life, but others remain ambulant into middle age and beyond.

CHARCOT-MARIE TOOTH DISEASE, ALSO KNOWN AS HEREDITARY MOTOR AND SENSORY NEUROPATHY OR PERONEAL MUSCULAR ATROPHY

Charcot-Marie-Tooth disease (CMT) is a group of neuromuscular diseases that have a variable inheritance pattern and vary in severity. It may first be noticed in childhood, affecting the small muscles of the hands (and fine motor movements) and feet (high arches, foot drop and 'club foot' may be symptoms).

Some people with the condition experience mild weakness and may not even be aware they have the condition, while others are severely affected and have great difficulty in walking.

CONGENITAL MUSCULAR DYSTROPHIES (CMD)

These are a collection of different muscular dystrophies characterised by weakness at infancy or early childhood. The severity of the condition depends on the type of congenital muscular dystrophy diagnosed. There are several different types of CMD which vary in symptoms, severity and progression. CMD affects both boys and girls. Symptoms include floppiness (hypotonia), poor head control, contractures, respiratory problems, swallowing and feeding difficulties. Some children may also have learning difficulties. While many children are never able to walk, others do achieve delayed walking but lose this ability as they grow older.

CONGENITAL MYOPATHIES

This group of conditions usually causes muscle weakness in children, although in some cases there are no symptoms until adulthood. Respiratory problems are a common feature and scoliosis, cardiac problems and contractures can also be factors, depending on the type of myopathy.

DUCHENNE MUSCULAR DYSTROPHY (DMD)

Duchenne muscular dystrophy is a serious condition and the most common of the childhood onset muscular dystrophies. It is caused by a fault on the X chromosome so the condition only affects boys with girls being carriers. The risk for the general population of having an affected child is 1:3500 births.

Duchenne is often, although not always, characterised by late walking (after 18 months). Further early signs include calf hypertrophy (enlarged calves) and muscle weakness in the lower limbs causing loss of balance. An affected child also finds it difficult to get up from the floor or use stairs. As the condition progresses, a distinctive walk emerges; to compensate for the increasing weakness in the hip and pelvic muscles the boy will

walk on his toes with his abdomen pushed forward. A child may also have contractures (stiffness) of the heels and ankles, which may require surgery.

Some boys with Duchenne have learning difficulties, particularly with language and communication skills. These difficulties are rarely severe and do not worsen over time.

Boys with Duchenne lose their ability to walk, usually after the age of nine, and then become full-time wheelchair users. From this point onwards, they may experience scoliosis (curvature of the spine), cardiac problems, weakness of the shoulders, arms and hands, chest infections and, at a later stage, respiratory problems. Life expectancy is reduced.

In the past, most boys died in their late teens. Today, with assisted non-invasive ventilation, they can survive into their late twenties and beyond, which poses new long-term management issues.

FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHY (FSHD)

FSHD is an inherited condition that can affect men and women. The first signs are usually weakness in the face and shoulder muscles which make it difficult for the individual to raise his or her arms. A 'winging' of the shoulder blades is also apparent. Weak facial muscles can affect speech, communication and feeding. The muscle which raises the foot is often affected early on in the condition - causing people to trip - along with the lower (distal) leg. This weakness can then spread to the larger hip girdle muscles. The rate at which the condition progresses is variable, although it is usually true that the earlier the symptoms, the more severe the eventual muscle problems. A minority of people will experience complete loss of walking, but others may also need a wheelchair for long distances and to prevent fatigue. Some people may have hearing loss.

INCLUSION BODY MYOSITIS

This is not an inherited condition. Inclusion body myositis is the most common form of muscle weakness acquired in later adult life. It causes substantial disability and is characterised by weakening muscles in the hands and thigh, which can cause falls. Swallowing may also be affected. Although other forms of myositis (muscle inflammation) respond to treatment, it is generally ineffective with this form of the condition.

LIMB GIRDLE MUSCULAR DYSTROPHIES

(LGMD) These are a group of progressive muscle conditions affecting both males and females. The limb girdle group of muscular dystrophies are so called because they usually cause weakness in the shoulder and pelvic girdle. Weakness in the legs generally occurs before weakness in the arms. The muscles of the face are usually unaffected. The condition progresses at a variable rate. Some children will be severely affected while others will not be diagnosed until adulthood. Some forms of limb girdle muscular dystrophies affect the heart and breathing. The course of LGMD is very variable and the rate of progression is difficult to predict.

METABOLIC MYOPATHIES

Contracting a muscle requires energy. The body metabolises the food eaten into a form of energy the muscle can use. Many inherited disorders affect the metabolic pathways. Some, such as McArdle's disease or mitochondrial myopathies, cause exercise intolerance (the person has no symptoms at rest but develops muscle pain and weakness if he or she attempts activity). Others, such as Pompe's Disease or debrancher enzyme deficiency, cause progressive weakness regardless of whether exercise is undertaken or not. Some metabolic myopathies also affect the heart and respiratory muscles.

MYOTONIC DYSTROPHY AND CONGENITAL MYOTONIC DYSTROPHY

Myotonic dystrophy and congenital myotonic dystrophy are dominantly inherited conditions that tend to increase in severity from one generation to the next. The majority of those affected will begin showing symptoms in early adulthood but it is not uncommon for children of affected mothers to have the more serious congenital form of the condition. Both men and women are equally likely to be affected.

A characteristic feature of these conditions is the myotonia or muscle stiffness (a delayed relaxation of the muscle following contraction) which is often worse in cold weather and more of a nuisance than a disability. More problematic, however, is muscle weakness in the hands, ankles, face and neck. Affected individuals may also experience smooth

muscle problems causing trouble with the gut (pain, similar to that experienced in Irritable Bowel syndrome, has been known to occur).

A significant feature of the conditions, particularly congenital myotonic dystrophy, is learning difficulties, which can be severe. People with myotonic dystrophy may also exhibit sleepiness; tiredness, lethargy and cataracts at an unusually early age are not uncommon. The heart may also be affected and cardiac monitoring is recommended for both adults and children. It is unlikely, however, that an individual would have all the symptoms and problems associated with such a variable and complex condition.

SPINAL MUSCULAR ATROPHY (SMA)

SMA is a recessively inherited condition that causes muscle weakness. It affects both boys and girls equally. The severity of the condition depends on the type of SMA and age of onset.

SMA Type I is the most severe form of the condition. Children with Type I are very weak and lack motor development. They cannot sit unaided and have difficulty breathing, sucking and swallowing. Most do not survive beyond their first birthday.

SMA with Respiratory Distress Type I (SMARD1) causes muscle weakness but the predominating symptom is severe respiratory distress due to paralysis of the diaphragm. Babies between one month and six months old experience respiratory failure and progressive muscle weakness, mainly in the distal lower limbs. Sensory and autonomic nervous systems may also be involved. SMARD1 is distinguishable from SMA Type I by the paralysis of the diaphragm and distal muscle weakness. Infants with SMA Type I become floppy due to weakness of the proximal limb muscles, assuming a 'frog leg' position, before they suffer respiratory failure due to paralysis of the intercostal muscles.

SMA Type II is slightly less severe than SMA Type I. Children with Type II can sit unaided and even stand with support, but cannot walk. They do not usually have difficulties with feeding and swallowing but are at increased risk of complications from respiratory infections. Some children will not live into adulthood.

SMA Type III is milder than SMA Type II. Children with Type III can stand and walk. They may outgrow their muscle strength and many do eventually need to use a wheelchair. SMA Type III affects children after 18 months of age.

SMA Type IV is the least common form of the condition and usually begins in late adolescence or adulthood. It has a similar clinical course to Becker muscular dystrophy and was often misdiagnosed as such in the past.

APPENDIX 2

CLINICAL CARE MANAGEMENT/CONSIDERATIONS

1. SPINAL PROBLEMS

In young children with conditions such as Spinal Muscular Atrophy Type II, some congenital muscular dystrophies, congenital myotonic dystrophy and some of the congenital myopathies, muscle weakness is present at or soon after birth. These children may begin to develop spinal curvature very early in life.

In clients with Duchenne muscular dystrophy (DMD), scoliosis often develops in the teenage years. The natural course of progression of scoliosis in DMD has been related to the type of spinal deformity[1]:

Type I — Scoliosis associated with kyphosis. This group demonstrates the greatest risk of rapid progression and is the most common pattern of spinal deformity in DMD.

Type II: — Scoliosis associated with hyper lordosis. This group is variable in its rate of progression.

Type III: — Stable.

The use of corticosteroids in the last two decades has changed the natural history of DMD. Corticosteroid use has been associated with the prolongation of ambulation,[2] improved cardiac and pulmonary function, and with a decrease in the incidence and severity of scoliosis.[3] [4] DMD boys on long-term steroid treatment are at increased risk of cataracts, weight gain, vertebral fractures, and long bone fractures.[4]

Management

Clients with neuromuscular conditions need to be followed by an orthopaedic surgeon to monitor spinal development and progression of spinal curvatures. Radiographs are warranted annually for curves of less than 15-20 degrees and every six months for curves of more than 20 degrees up to skeletal maturity.[5]

Spinal bracing

There is currently no evidence that bracing reduces the rate of progression of the scoliosis.[6] The primary indication for use of bracing is as an adjunct to seating to assist with postural control. It must, however, be considered that rigid spinal bracing may compromise respiratory function so this intervention must be used with caution.[7] [8]

Spinal surgery

The right time to perform surgery needs to be decided in consultation between the young person and his or her family, the spinal surgeon, the neuromuscular team and the anesthetist. The possibility of surgery needs to be considered in very close relation to the child's respiratory and cardiac function.

It is not usually necessary to consider surgery in very young children when years of rapid spinal growth lie ahead, unless the deformity is severe. However, some children with SMA, or congenital conditions, may require surgery at a young age (this can be as young as four or five-years-old). More than one operation may be needed to accommodate the child's growth.

---

[1] Oda T, Shimizu N, Yonenobu K, et al. Longitudinal study of spinal deformity in Duchenne muscular dystrophy. *Journal of Pediatric Orthopedics.* 1993; 13: 478-488.

[2] King WM, Ruttencutter R, Nagaraja HN, Matkovic V, Landoll J, Hoyle C, Mendell JR, Kissel JT. Orthopedic outcomes of long-term daily corticosteroid treatment in Duchenne muscular dystrophy. *Neurology.* 2007; 58: 1607-1613.

[3] Alman BA, Raza SN, Biggar WD. Steroid treatment and the development of scoliosis in males with Duchenne Muscular Dystrophy. The *Journal of Bone and Joint Surgery.* 2004; 86(3): 519-524.

[4] Biggar WD, Harris VA, Eliasoph L, Alman B. Long-term benefits of deflazacort treatment for boys with duchenne muscular dystrophy in their second decade. *Neuromuscular Disorders.* 2006; 16: 249-255.

[5] Bushby K, Finkel R, Birnkrant DJ, Case LE, Clemens PR, et al. Diagnosis and management of Duchenne muscular dystrophy, part 2: implementation of multidisciplinary care. *Lancet Neurology.* 2010; 9: 177-89.

[6] Mullender MG, Blom NA, De Kleuver M, Fock JM, et al. A Dutch guideline for the treatment of scoliosis in neuromuscular disorders. *Scoliosis.* 2008; 3(14): doi:10.1186/1748-7161-3-14.

[7] Noble-Jamieson CM, Heckmatt JZ, Dubowitz V, Silverman M. Effects of posture and spinal bracing on respiratory function in neuromuscular disease. *Archives of Disease in childhood.* 1986; 61: 178-181.

[8] Tangsrud SE, Carlsen KC, Lund-Petersen I, Carlsen KH. Lung function measurements in young children with spinal muscle atrophy: a cross sectional survey on the effect of position and bracing. *Archives of Disease in Childhood.* 2001; 84: 521-524.

Following spinal instrumentation surgery clients will no longer be able to lean forward to complete functional tasks. This can be a problem for clients with neuromuscular conditions who are accustomed to leaning forward in order to bring their mouth closer to their hands to eat independently, or to play video games and access other technology. This can be emotionally devastating for clients and for caregivers. A discussion prior to surgery about alternate strategies or equipment that will be required post-operatively to assist with functional tasks can help make the adjustment to the new posture easier.

It will be *essential* for the spinal team, therapists and the seating service therapists to liaise prior to surgery to discuss the user's post-operative wheelchair and seating requirements, and any alterations or additional features that may be needed.

2. RESPIRATORY MANAGEMENT/DAY TIME VENTILATION

Many different neuromuscular conditions cause weakness of the breathing muscles. This can affect both children and adults at any age. Regular monitoring of respiratory function by a Respirologist is essential. As respiratory function deteriorates, clients with neuromuscular conditions may require non-invasive positive pressure ventilation at night. Many people progress to requiring non-invasive positive pressure ventilation during the day either intermittently or permanently. Occasionally, a person with a neuromuscular condition will be ventilated via a tracheostomy. The wheelchair must be able to accommodate all the essential ventilation equipment.

Ventilation equipment considerations:

- Ventilator (consideration of type, size, access, power supply)
- Possible need for humidifier, back-up battery, suction machine, oxygen, bag and mask, spare tubing, suction catheters, etc.
- Tilt testing of chair, user and all equipment

APPENDIX 3

TRANSPORTATION

SelfCare

Buses of the Lower Mainland

May 2011

Vancouver 604-872-5800 / North Shore 604-990-9422 / Sechelt 604-741-9920 / Surrey 604-574-5801

Trolley Bus *Used in Vancouver City only* • Called a Lo-Floor • Comes in 40' and 60' configurations • Not the narrowest ... great for scooters not larger than the 2' X 4' template • Pretty good access past the farebox • Has the 'non-securement' area	
Orion or High Platform Bus *Used in Richmond only* • Called a Hi-Floor • Used primarily as a highway commuter bus, in and out of Richmond • Uses a 'LIFT SYSTEM'...the whole lift is lowered, the customer drives the scooter/chair on to the lift, then the Operator operates the lift up in to the coach	
Flyer Bus *Used in Surrey, Port Coquitlam, Burnaby* • Called a Lo-Floor 'Vancouver-style'40' Coach because it looks like the Trolley Coach - is a Conventional 40' diesel bus • Uses a 'ramp system' that is deployed after the coach is 'lowered' to make the floor of the coach even with the curb • Does not have the 'non-securement' area • Scooters and wheelchairs must be secured by strapping the scooter down	

Terms used:
Non-securement: Scooters are parked facing the rear of the bus and are NOT strapped into the bus.
Ramp system: An inclined ramp is deployed for the rider to drive up and into the bus.
Lift system: The rider drives onto the lift and the operator lifts the rider into the bus.

Nova Bus *Used in Vancouver, Burnaby, North Vancouver* • Lo-Floor • Newest bus with widest entry (an extra 1/3 of the front door opens, to allow ease of access) • Uses a 'ramp system' • Has one scooter/wheelchair 'non-securement' area	
Flyer Lo-Floor Bus *Used in all locations* • One of the very first Lo-Floor configurations in the GVRD • Uses 'ramp-system' but does not have a 'non-securement' area (all scooters are strapped in) • Customers using this coach get used to the 'snug' entry passing the farebox	
Community Shuttle Coach *Used in all locations* • Uses a 'LIFT-system' for delivering customers using scooters and wheelchairs in to the coach, customers just need to back onto the ramp that lowers from the side of the coach, near the rear • Secured in the two locations available on the coach • Will take a bit larger scooter (Pegasus3)	

Terms used:
Non-securement: Scooters are parked facing the rear of the bus and are NOT strapped into the bus.
Ramp system: An inclined ramp is deployed for the rider to drive up and into the bus.
Lift system: The rider drives onto the lift and the operator lifts the rider into the bus.

Before using a scooter on the bus, contact your local transit depot to arrange training. Coast Mountain Bus Company has excellent trainers who are willing to spend the time to help scooter or power wheelchair users practice on parked buses that are used in their area. Contact your local SelfCare Seating & Mobility Consultant to trial a scooter on the bus before making a purchase decision.

**This listing is for informational purposes only and represents the express opinions of SelfCare Home Health Products Ltd. It does not necessarily reflect the views of Coast Mountain Bus Company. SelfCare assumes no liability for any inaccurate, delayed or incomplete information, or for any actions taken in reliance thereon.*

Special Recommendations for:

Name_____:

_____

_____

_____

Disclaimer

The following information, i.e. guideline/educational material/policy or procedure has been developed for use only within Sunny Hill Health Centre for Children (Sunny Hill). There are support systems at Sunny Hill that may not exist in other clinical setting, and therefore, any adoption of these materials cannot be the responsibility of Sunny Hill. Agencies other than Sunny Hill should use this information as a guideline for reference purposes only. All materials are the property of Sunny Hill and may only be reprinted in whole or in part with our expressed permission.

Your therapist's name: _____

Contact telephone number: _____

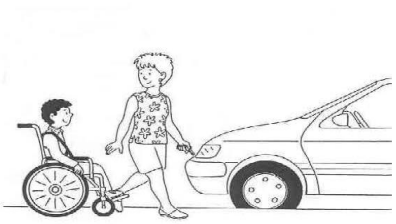
Date: _____

created April 2002 last update: July 2012

SUNNY HILL HEALTH CENTRE FOR CHILDREN
part of Children's & Women's Health Centre of British Columbia

3644 Slocan Street
Vancouver, BC V5M 3E8

Wheelchair Transportation "Safety Tips" for Children and Youth



Basic Principle

* The safest place to be transported is on the vehicle seat using the vehicle seatbelt system. Those who can transfer and ride safely on the vehicle seat or commercial car seat should do so.

DO's

√ All wheelchairs should face toward the front of the vehicle.

√ Always have the wheel locks on.

√ Power wheelchairs must be turned "off".

√ If your child has poor head control, a soft neck collar may be recommended. (Please consult your therapist).

√ Use a wheelchair tie-down restraint system to identified tie down locations on the wheelchair frame

√ Secure the passenger with a shoulder and lap belt.

√ The lap belt should fit over the hips not the stomach.

√ Securely store all loose items in the vehicle (including tray, medical equipment etc.).

√ Use a headrest.

DO NOT's

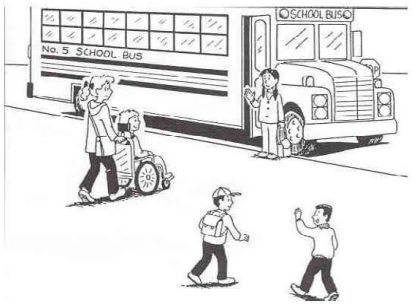
∅ Do NOT transport the wheelchair in a tilted or reclined position unless medically required and then only up to 30 degrees from upright.

∅ Do NOT use the tray during transport unless it is made of foam.

∅ Do NOT use head straps during transport.

∅ Do NOT attach neck collars to the wheelchair or seating system.

∅ Do NOT fasten tie-down straps to movable parts of the wheelchair such as footrests, wheels or front hangers.



TRANSPORTATION RESOURCE LINKS

There are a variety of resources available regarding transportation of individuals in wheelchairs:

BC Motor Vehicle Act
Motor Vehicle Restraint Systems and Booster Seats Safety Regulations (SOR/2010-90)
Child Seating and Restraint Systems

RESNA (Rehabilitative Engineering and Assistive Technology Society of North America
RESNA's Position on Wheelchairs Used as Seats in Motor Vehicles

Companies providing vehicle modifications:

Lower Mainland:
- Sidewinder (Chilliwack)
- Shoppers Home Health (Burnaby)
- Motion Specialties (Vancouver)

Prince George
- PG Surg-Med Ltd.

Vancouver Island
- Shoppers Home Health (Victoria)
- Motion Specialties (Victoria and Nanaimo)

Okanagan
- Surg-Med (Vernon)
- Can Am Mobility (Kelowna)

PUBLIC TRANSPORTATION

Translink: Access Transit Secretariat

Disabled World: Accessible Vancouver

Rehabilitation Engineering Research Center (RERC) on Wheelchair Transportation Safety
Transportation Review Checklist
This checklist is a model for reviewing the safety issues that affect children who ride in wheelchairs on school buses. (2007)

GENERAL RESOURCES

Guidelines for Use of Secondary Postural Support Devices by Wheelchair Users during Travel in Motor Vehicles

AIR TRANSPORTATION

Canadian Transportation Agency: Accessible Transportation

Air Canada: Customers with special needs

WestJet: Guests with special needs

SCHOOL BUS TRANSPORTATION

School District # _____

Student Name: _____ Date of Birth: _____

School: _____ Weight: _____

Date: _____ Height: _____

Who is the Case Manager and/or Resource Teacher(s) and/or Therapist?

Name(s): _____ / _____

Role(s): _____ / _____

Contact number(s): _____ / _____

Who is aware of the transportation plan?

Driver _____ Yes ☐ No ☐ n/a ☐

Education Assistant _____ Yes ☐ No ☐ n/a ☐

Resource Teacher _____ Yes ☐ No ☐ n/a ☐

Principal _____ Yes ☐ No ☐ n/a ☐

Therapist (OT or PT) _____ Yes ☐ No ☐ n/a ☐

Caregiver/Parents _____ Yes ☐ No ☐

(Please note: Law, Best Practice and Notes below are in italic)

1. Vehicle(s) for Transportation Bus ☐ Taxi ☐ Caregiver/Parent vehicle ☐

Comments:

Law: *To sit on the vehicle seat a student must be at least 9 years old or over 4'9" (145 cm) tall [BC Motor Vehicle Act Regulation Division 36 (Division 36)].*

Note: *If the student's ability to transfer into the vehicle, onto the seat or sit independently is an issue, please contact the therapist working with that student for a consultation.*

2. Will the student sit on the vehicle seat? Yes ☐ No ☐

Comments:

3. Will the student be transported in a safety seat or booster seat?
If "Yes," please comment on the process. If "No," consult Division 36 for legal requirements. Yes ☐ No ☐

Comments:

Law: *Taxis are exempt from Division 36 Child Seating and Restraint Systems, taxis are not legally required to use child safety seats.*
Best Practice is to keep children as safe as possible and use of child safety seats in taxis is always recommended.

4. If the student is transported in a wheelchair:
STUDENT'S EQUIPMENT NEEDS:

A. Head rest — Yes ☐ No ☐

Best Practice: Headrests are almost always recommended.

B. Positioning straps — Yes ☐ No ☐

Tick all the straps used when transported — Shoulder ☐ Chest ☐ Pelvic ☐ Shin☐ Feet ☐ Arms ☐

Note: Positioning straps are not crash tested and do not replace a seatbelt attached to the vehicle frame.

C. Foam cervical neck collar — Yes ☐ No ☐

Best Practice: Only soft foam collars, if needed, are recommended.

D. Specific student needs i.e., vents, special neck collars, foam trays, buckle guards, oxygen tanks, suction or feeding machines etc. *Make notes below.*

WHEELCHAIR SET UP:

A. Is the wheelchair:

Upright? (if "NO" specify the tilt degree) — Yes ☐ No ☐ Tilt degree _____

Tilted? This student needs _____ degrees tilt

Best Practice: up to 30 degrees is accepted — Yes ☐ No ☐ n/a ☐

B. Are the anti-tippers on? — Yes ☐ No ☐

C. Identified tie down locations on the wheelchair? — Yes ☐ No ☐

Best Practice: Many wheelchairs have a "D" -shaped brackets specific for tie down hooks. If the "D"-shaped brackets are not present attach the hooks to the frame of the wheelchair, avoiding any moving parts such as the footrest hanger or wheels. If you are not sure, ask the student's therapist or wheelchair dealer. Most wheelchairs have the dealer's contact information sticker on the wheelchair.

Comments:

VEHICLE SET UP:

A. Tie downs — Yes ☐ No ☐

B. Seatbelt — Yes ☐ No ☐

Tick what is used: — Lap belt ☐ Shoulder/Lap belt ☐

Note: Shoulder belts should be used if available and fit across the shoulder. Lap belts are required by law, see below.
Law: *All taxis are required to have tie downs and a seatbelt (see BC Motor Vehicle Act Regulations Division 44) and it is the driver's responsibility to apply the seatbelt or in the case of minors, assist the caregiver to put on the seatbelt. School buses are held to the standard at the date of manufacture, so some older school buses that are wheelchair equipped do not have a shoulder/lap belt system. If this is the case a lap belt with a metal buckle attached to wheelchair frame is recommended.* **Is this in place? Yes ☐ No ☐ N/A ☐**

Comments:

DURING TRAVEL:

The following items, if used by the student, need to be removed and stored securely during travel:

A. Rigid tray — Yes ☐ No ☐

B. Head bad attached to the headrest — Yes ☐ No ☐

C. Communication or computer system and mount — Yes ☐ No ☐

Comments:

APPENDIX 4

HIGH PERFORMANCE MANUAL WHEELCHAIRS

There are now many high performance manual wheelchairs available. The frames are rigid or folding and made from a variety of materials. Seat width, seat depth and backrest height can be specified to meet an individual's needs and most manufacturers provide a range of accessories. The rear wheels are usually 'quick release' and adjust up and down in the frame, to change seat height and angle. They can also move backwards and forwards to adjust the balance position. (Castor housings and forks adjust to accommodate the movement of the rear wheel.)

Photo: Tilite.com

Benefits of this type of wheelchair for people with neuromuscular conditions:

CUSTOM FIT

The choice of seat width, height and depth, height of the backrest, armrest and footrest ensures that the wheelchair fits the user. A good fit promotes good postural support and discourages postures which could cause deformities to develop.

EASE OF PROPULSION/CONTROL

The positioning of the rear wheels and the use of lightweight materials make these wheelchairs easier to push and maneuver than standard manual wheelchairs. The rear wheels are positioned in front of the back rest which enables the user to easily perform a 'back wheel balance' that aids independent use and control. Low profile, high-pressure tires are also beneficial.

SEAT HEIGHT

The seat height can be increased by moving the rear wheels down the frame and adjusting the front fork and castor housing assembly. Increased seat height can help users who are still able to stand. Full-length armrests are also beneficial. A careful assessment of the way the user moves from sitting to standing will be needed to help decide the choice of frame style and footplate.

INCREASED INDEPENDENCE

Increasing the ability to self-propel may reduce the need for the user to be accompanied and pushed. The light frame and quick release wheels may also mean that some users can lift the wheelchair into, and out of, their car.

EXERCISE

Even if a user is no longer able to walk, self-propulsion ensures that he or she is undertaking some physical activity. This benefits general health and assists with weight control. This type of wheelchair can encourage active use of the arms which may help maintain muscle strength.

ASSESSMENT

Representatives from most companies selling this type of wheelchair can usually attend assessments and provide information on the technical specifications of the wheelchair. While many of these representatives are very experienced at assessments, they are unlikely to know about the needs of users with neuromuscular conditions. It is recommended that a suitably experienced therapist is present at any assessment.

High performance manual wheelchairs are under-issued for users with neuromuscular conditions, possibly because the criteria of many Wheelchair Services exclude the provision of this type of wheelchair to anyone other than a full-time user. This type of wheelchair can benefit users with a neuromuscular condition by improving health, slowing deterioration and increasing independence.

APPENDIX 5

FUNDING OPTIONS FOR MOBILITY EQUIPMENT IN BRITISH COLUMBIA

There are a variety of programs and funding sources in British Columbia for people with neuromuscular conditions which include Ministry and Health Canada funded government programs, private pay coverage, and disability or community organizations. Seating therapists work with social workers to assist clients in accessing appropriate programs. Often equipment is partially funded through a combination of funding options.

Ministry & Health Canada Programs

1. **The Ministry of Children and Family Development** At Home Program provides funding for basic mobility equipment and seating systems.

   Eligibility:

   - Age: 17 years or younger for Medical Benefits;
   - A resident of British Columbia;
   - Enrolled with British Columbia Medical Services Plan;
   - Living at home with a parent or guardian, and;
   - Assessed as dependent in at least three of the four activities of daily living (eating, dressing, toileting and washing)

   - At Home Program Guide
   - At Home Program Medical Benefits Transition to Disability Assistance- July 2013
   - 17-Year Old Disability Assistance Applicants

2. **Ministry of Children and Family Development** provides equipment funding for children and youth in foster care with special needs. The Health Supports for Children in Care and Youth Agreements details the coverage for medical equipment funding.

3. **Ministry of Social Development and Social Innovation**

   Eligibility: Medical equipment and devices for medically essential needs are available to adult clients who are eligible for general health supplements.

   - Medical Equipment and Devices - Overview
   - Medical Equipment and Devices - Policy
   - Medical Equipment and Devices - Procedures
   - Medical Equipment and Devices - Forms and Letters
   - Medical Equipment and Devices - Authorities and Responsibilities
   - Contact a Ministry Worker/Social Development and Social Innovation Office Directory
   - Persons with Disabilities (PWD) Designation

Must contact Health Assistance Branch when:

- Assessing eligibility for equipment rentals for more than 60 days
- Assessing eligibility for equipment purchases and repairs over $500

4. The **BC Government** Personal Supports Directory describes additional programs for adults who may be eligible for mobility equipment funding:

   - Employment and Income Assistance for Persons with Disabilities
     Eligibility: Employment and Assistance client
   - Employment Program of British Columbia (EPBC)
     Eligibility: Individuals who have a disability that is the primary barrier to finding and maintaining paid or volunteer employment
   - Equipment and Assistive Technology Initiative (EATI)
     Eligibility: People with disabilities who would like to work

5. Health Canada Non-Insured Health Benefits Program offers coverage for medical mobility equipment.
   Eligibility: Eligible First Nations and Inuit who have a status and band number

Private Pay Options

Clients may have access to:

1. Extended Health Plan Coverage
2. Private Insurance Plans
3. Private funds

Therapists, social workers and seating clinics may be aware of additional local disability groups, funding programs or community organizations that can assist with funding for mobility equipment.

Disability Groups

Muscular Dystrophy Canada Western Region Equipment Program

Community Organizations

Muscular Dystrophy Canada provides an Equipment Funding Resource Guide that can assist individuals in finding alternate or additional funding for medical equipment.

Muscular Dystrophy Canada's Safeway Mobility Grants.

EVIDENCE TABLES FOR INTERVENTION STUDIES

CHAPTER 2: PRE-SCHOOL-AGED CHILDREN

Citation	Study Design	Level of Evidence	Study Objective/Purpose	Population	Key Clinical Messages
Butler C, Okamoto GA, McKay TM. Powered mobility for very young disabled children. *Developmental Medicine & Child Neurology.* 1983; 25(4): 472-4.	D/CS	5	Can young children use a power wheelchair?	9 children aged 20-39 months with physical disabilities including Spinal Muscular Atrophy (SMA).	Young with physical but no cognitive disability can learn to drive safely and independently within a very short period of time.
Butler C, Okamoto GA, McKay TM. Motorized wheelchair driving by disabled children. *Archives of Physical Medicine & Rehabilitation.* 1984; 65(2): 95-7.	D/CS	5	Is competent control of a power wheelchair attainable by young children?	13 children aged 20-37 months with physical disabilities including SMA.	Children as young as 24 months can learn to drive powered wheelchairs. Start/stop and circling appear to be the most common skills that children learn first.
Dunaway S. et al (2012). Independent mobility after early introduction of a power wheelchair in spinal muscular atrophy. *Journal of Child Neurology.* 2013; 28(5): 576-82.	D/CS	5	Explore the feasibility of independent mobility in young children with SMA.	4 children aged 24-34 months (at wheelchair delivery) with SMA or congenital MD.	Young children with SMA can achieve independent use of a power wheelchair and this should be introduced between 7 and 24 months of age.
Jones MA, McEwen IR, Hansen L. Use of power mobility for a young child with Spinal Muscular Atrophy. *Physical Therapy.* 2003; 83(3): 253-62.	CS	5	Demonstrate that a child as young as 20 months can use a power chair competently. To evaluate developmental change after provision of a power wheelchair.	A 20-month old child with SMA.	Provision of power mobility can increase overall independence. Power mobility use may be associated with developmental change.
Guerette P, Furumasu J, Tefft D. The positive effects of early powered mobility on children's psychosocial and play skills. *Assistive Technology.* 2013; 25 (1): 39-48.	B&AC	4	Explore impact on children's psychosocial and play skills of the early introduction of power mobility.	10 children aged 18-42 months with physical disabilities including SMA and 13 aged 18-72 months with CP.	Power mobility can impact positively on children's social and play skills as well as increasing level of independence in mobility activities.
Tefft D, Guerette P, Furumasu J. The impact of early powered mobility on parental stress, negative emotions and family interactions. *Physical and Occupational Therapy in Pediatrics.* 2011; 31(1); 4-15.	B&AC	4	To explore the impact on families of the introduction of power mobility.	10 children aged 18-42 months with physical disabilities including SMA and 13 aged 18-72 months with CP.	Introduction of a powered mobility device may improve parents' satisfaction with their child's play, social skills, sleep/wake cycles and level of independence.

B&AC Before and After Cohort
CS Case Study
D/CS Descriptive/Case Studies

CHAPTER 3: SCHOOLCHILDREN TO ADOLESCENTS

Citation	Study Design	Level of Evidence	Study Objective/Purpose	Population	Key Clinical Messages
Clark J, Michael S, Morrow, M. Wheelchair postural support for young people with progressive neuromuscular disorders. *International Journal of Therapy and Rehabilitation.* 2004; 11(8): 356-371.	B&ACS	4	Evaluate effects of postural supports on posture, respiration and upper limb function.	4 boys with Friedreich's Ataxia and 15 boys with DMD. Aged 6-21 yrs.	Adaptive seating can change body alignment. Adaptive seating should be used within the first wheelchair to help minimize postural asymmetry. No immediate effects on respiratory or upper limb function were found in this study.
Evans S, Neophytou L, De Souza L, Frank A. Young people's experiences using electric powered indoor-outdoor wheelchairs (epiocs) : Potential for enhancing users' development ? *Disability and Rehabilitation.* 2007; 29(16): 1281-1294.	PH	N/A	Describe experiences of severely physically disabled young people who use electric powered indoor-outdoor wheelchairs (EPIOCs).	18 young people including 10 with muscular dystrophy. Aged 10-19 yrs.	Important to provide a manual wheelchair that is not just a 'back-up.' Additional wheelchair skills and safety training may be important at this age. Increased suspension in the wheelchair is needed to alleviate pain and discomfort. Swing away joysticks allow access to tables. Consider wheelchair features necessary for participation in sports such as Power Soccer.
Flodin E. Interactive design - the desire for autonomous upright mobility: A longitudinal case study. *Technology and Disability.* 2007; 19: 213-224.	CS	5	Describe the process and evolution of a custom upright powered mobility device for one child.	1 girl with SMA type II from age 1 year 5 months to 12 years 11 months.	Young children can participate in making choices about their own positioning and mobility equipment. Independent mobility in an upright position may offer psychological and physical benefits for children with neuromuscular conditions.
Kerr TP, Lin JP, Gresty MA, Morley T, Robb SA. Spinal stability is improved by inducing a lumbar lordosis in boys with Duchenne Muscular Dystrophy: A pilot study. *Gait & Posture.* 2008; 28:108-112.	B&ACS	4	To compare ability to tolerate lateral trunk loading when seated on a flat seat versus seated on a 15 degree anterior wedge.	8 boys with DMD and no scoliosis, aged 8.1-11.1 years. 10 typically developing boys aged 7.2-14.4 years.	For boys with DMD who can tolerate this position, a 15 degree anterior wedge encourages a lumbar lordosis. In boys, who do not already have a scoliosis, this position may increase ability to tolerate lateral trunk loading. The authors conclude that this position may help

Citation	Study Design	Level of Evidence	Study Objective/Purpose	Population	Key Clinical Messages
					prevent or delay progression of a scoliosis. Whether external force application relates to scoliosis progression is questionable.
Liu M, Mindeo K, Hanayama K, Fujiwara T, Chino N. Practical problems and management of seating through the clinical stages of Duchenne's Muscular Dystrophy. *Archives of Physical Medicine and Rehabilitation.* 2003; 84: 818-824.	CR-S	5	Describe seating problems in patients with DMD.	95 individuals with DMD aged 8-33 yrs. 93 male. Mean age 15.9	Initial seating support should aim to keep pelvis level and lower limbs symmetrical. Many clients with DMD have pain and pressure issues. Trunk support should be customized to help counteract spinal deformities while facilitating function.
Richardson M, Frank AO. Electric powered wheelchairs for those with muscular dystrophy: Problems of posture, pain and deformity. *Disability and Rehabilitation: Assistive Technology.* 2009; 4(3): 181-188.	CR-S + RCR	5	To identify issues of posture, pain and deformity in clients with MD.	29 individuals with MD aged 10-63.	Need frequent seating and mobility reviews. Provision of postural support in the first wheelchair. Tilt and recline to alleviate pain and discomfort. Programmable electronics to accommodate changing needs.
Wong C, Wade C. Reducing iliotibial band contractures in patients with muscular dystrophy using custom dry floatation cushions. *Archives of Physical Medicine and Rehabilitation.* 1995; 76: 695-700.	B&ACS	4	Evaluate use of custom dry floatation cushions reduce ITB contractures and provide adequate comfort.	9 clients with DMD or limb girdle muscular dystrophy aged 9-69 years.	Lateral thigh support can help reduce and prevent development of ITB contractures. Custom dry floatation cushions can be used to provide this level of support.

B&ACS Before and After Case Series
CS Case Study
CR-S Cross-sectional
PH Phenomenology
RCR Retrospective Chart Review

CHAPTER 4: ADOLESCENTS TO YOUNG ADULTS

Citation	Study Design	Level of Evidence	Study Objective/Purpose	Population	Key Clinical Messages
Clark J, Michael S, Morrow M. Wheelchair postural support for young people with progressive neuromuscular disorders. *International Journal of Therapy and Rehabilitation.* 2004; 11(8): 356-371.	B&ACS	4	Evaluate effects of postural supports on posture, respiration and upper limb function.	4 boys with Friedreich's Ataxia and 15 boys with DMD. Aged 6-21 yrs.	Adaptive seating can change body alignment. Adaptive seating should be used within the first wheelchair to help minimize postural asymmetry. No immediate effects on respiratory or upper limb function were found in this study.
Evans S, Neophytou L, De Souza L, Frank A. Young people's experiences using electric powered indoor-outdoor wheelchairs (epiocs) : Potential for enhancing users' development ? *Disability and Rehabilitation.* 2007; 29(16): 1281-1294.	PH	N/A	Describe experiences of severely physically disabled young people who use electric powered indoor-outdoor wheelchairs (EPIOCs).	18 young people including 10 with muscular dystrophy. Aged 10-19 yrs.	Important to provide a manual wheelchair that is not just a 'back-up.' Additional wheelchair skills and safety training may be important at this age. Increased suspension in the wheelchair is needed to alleviate pain and discomfort. Swing away joysticks allow access to tables. Consider wheelchair features necessary for participation in sports such as Power Soccer.
Liu M, Mindeo K, Hanayama K, Fujiwara T, Chino N. Practical problems and management of seating through the clinical stages of Duchenne's Muscular Dystrophy. *Archives of Physical Medicine and Rehabilitation.* 2003; 84: 818-824.	CR-S	5	Describe seating problems in patients with DMD.	95 individuals with DMD aged 8-33 yrs. 93 male. Mean age 15.9.	Initial seating support should aim to keep pelvis level and lower limbs symmetrical. Many clients with DMD have pain and pressure issues. Trunk support should be customized to help counteract spinal deformities while facilitating function.
Richardson M, Frank AO. Electric powered wheelchairs for those with muscular dystrophy: Problems of posture, pain and deformity. *Disability and Rehabilitation: Assistive Technology.* 2009; 4(3): 181-188.	CR-S + RCR	5	To identify issues of posture, pain and deformity in clients with MD.	29 individuals with MD aged 10-63.	Need frequent seating and mobility reviews. Provision of postural support in the first wheelchair. Tilt and recline to alleviate pain and discomfort. Programmable electronics to accommodate changing needs.

Citation	Study Design	Level of Evidence	Study Objective/Purpose	Population	Key Clinical Messages
Wong C, Wade C. Reducing iliotibial band contractures in patients with muscular dystrophy using custom dry floatation cushions. *Archives of Physical Medicine and Rehabilitation.* 1995; 76: 695-700.	B&ACS	4	Evaluate use of custom dry floatation cushions reduce ITB contractures and provide adequate comfort.	9 clients with DMD or limb girdle muscular dystrophy aged 9-69 years.	Lateral thigh support can help reduce and prevent development of ITB contractures. Custom dry floatation cushions can be used to provide this level of support.

B&ACS Before and After Case Series
CR-S Cross-sectional
PH Phenomenology
RCR Retrospective Chart Review

CHAPTER 5: ADULTS

Citation	Study Design	Level of Evidence	Study Objective/Purpose	Population	Key Clinical Messages
Clark J, Michael S, Morrow M. Wheelchair postural support for young people with progressive neuromuscular disorders. *International Journal of Therapy and Rehabilitation.* 2004; 11(8): 356-371.	B&ACS	4	Evaluate effects of postural supports on posture, respiration and upper limb function.	4 boys with Friedreich's Ataxia and 15 boys with DMD. Aged 6-21 yrs.	Adaptive seating can change body alignment. Adaptive seating should be used within the first wheelchair to help minimize postural asymmetry. No immediate effects on respiratory or upper limb function were found in this study.
Liu M, Mindeo K, Hanayama K, Fujiwara T, Chino N. Practical problems and management of seating through the clinical stages of Duchenne's Muscular Dystrophy. *Archives of Physical Medicine and Rehabilitation.* 2003. 84: 818-824.	CR-S	5	Describe seating problems in patients with DMD.	95 individuals with DMD aged 8-33 yrs. 93 male. Mean age 15.9.	Initial seating support should aim to keep pelvis level and lower limbs symmetrical. Many clients with DMD have pain and pressure issues. Trunk support should be customized to help counteract spinal deformities while facilitating function.
Pellegrini N, Guillon B, Prigent H, et al. Optimization of power wheelchair control for patients with severe Duchenne Muscular Dystrophy. *Neuromuscular Disorders.* 2004; 14(5): 297-300.	CR-S	5	To investigate the relationship between age, upper limb strength and ability to use a standard joystick without fatiguing.	84 individuals with DMD aged 25.3 years ±5 years.	Key pinch strength is associated with ability to use a standard joystick. Clients with DMD progressively lose ability to use a standard joystick with increasing age but can regain driving independence through use of alternate controls such as a mini joystick
Richardson, M, Frank, AO. Electric powered wheelchairs for those with muscular dystrophy: Problems of posture, pain and deformity. *Disability and Rehabilitation: Assistive Technology.* 2009; 4(3): 181-188.	CR-S + RCR	5	To identify issues of posture, pain and deformity in clients with MD.	29 individuals with MD aged 10-63.	Need frequent seating and mobility reviews. Provision of postural support in the first wheelchair. Tilt and recline to alleviate pain and discomfort. Programmable electronics to accommodate changing needs.

Citation	Study Design	Level of Evidence	Study Objective/Purpose	Population	Key Clinical Messages
Wong C, Wade C. Reducing iliotibial band contractures in patients with muscular dystrophy using custom dry floatation cushions. *Archives of Physical Medicine and Rehabilitation.* 1995; 76: 695-700.	B&ACS	4	Evaluate use of custom dry floatation cushions reduce ITB contractures and provide adequate comfort.	9 clients with DMD or limb girdle muscular dystrophy aged 9-69 years.	Lateral thigh support can help reduce and prevent development of ITB contractures. Custom dry floatation cushions can be used to provide this level of support.

B&ACS Before and After Case Series
CR-S Cross-sectional
RCR Retrospective Chart Review

WEB REFERENCES

Child Development & Rehabilitation	http://www.childdevelopment.ca
(UK) Muscular Dystrophy Campaign	http://www.muscular-dystrophy.org
Air Canada: Customers with special needs	http://www.aircanada.com/en/travelinfo/before/specialneeds.html
BC Housing: Home Adaptations for Independence	http://www.bchousing.org/Options/Home_Renovations
Body Measurement chart	http://sh-elearn.atutor.ca/go.php/17/content.php/cid/380/
Canada Mortgage and Housing Corporation: Accessible Housing by Design	http://www.cmhc-schl.gc.ca/en/co/acho/acho_001.cfm
Canadian Transportation Agency: Accessible Transportation	http://www.cta-otc.gc.ca/eng/accessible-transportation
Disabled World: Accessible Vancouver	http://www.disabled-world.com/travel/canada/britishcolumbia/vancouver.php
Driver Rehab	http://www.vch.ca/media/GFS_ABIResourceDirectory.pdf
GF Strong Rehabilitation Centre	http://www.vch.ca/EN/find_locations/find_locations/?&site_id=73
GF Strong Rehab Centre: Assistive Technology & Seating Service	http://www.assistive-technology.ca/deal.html
Guidelines for Use of Secondary Postural Support Devices by Wheelchair Users during Travel in Motor Vehicles	http://www.rercwts.pitt.edu/RERC_WTS2_KT/RERC_WTS2_KT_Pub/RERC_WTS_Pub_Doc/RERC_WTS_032_06.pdf
Health Canada Non-Insured Health Benefits Program	http://www.hc-sc.gc.ca/fniah-spnia/nihb-ssna/benefit-prestation/medequip/index-eng.php
HealthLink BC	http://www.healthlinkbc.ca/
Muscular Dystrophy Canada	http://muscle.ca/
Muscular Dystrophy Canada: Equipment Funding Resource Guide	http://www.muscle.ca/fileadmin/Western/Services/Equipment_Funding_Manual-WC-Aug07-2009.pdf
Muscular Dystrophy Canada: Western Region Equipment Program	http://www.muscle.ca/fileadmin/Western/Services/mobility_equipment_program.pdf
Muscular Dystrophy Canada's Safeway Mobility Grants	http://www.muscle.ca/fileadmin/Western/Fundraising/Campaigns_Events/Safeway/Safeway_Guidelines_and_Grant_Application_Form_2011.pdf
Persons with Disabilities (PWD) designation	http://www.gov.bc.ca/meia/online_resource/health_supplements_and_programs/healthsuppsum/definitions.html#12
Positioning and Mobility worksheet	http://sh-elearn.atutor.ca/go.php/20/content.php/cid/383/ib/1/p_course/20
Postural Problems and Solutions chart	http://sh-elearn.atutor.ca/go.php/18/content.php/cid/382/
Range of Motion chart	http://sh-elearn.atutor.ca/go.php/16/content.php/cid/150/
Red Cross Children's Medical Equipment Recycling and Loan Service (CMERLS)	http://www.redcross.ca/where-we-work/in-canada/british-columbia-and-yukon/bc-specific-programs/children-s-medical-equipment-recycling-and-loan-service
Rehabilitation Engineering Research Center (RERC) on Wheelchair Transportation Safety: Transportation Review Checklist	http://www.rercwts.org/rerc_wts2_kt/rerc_wts2_kt_stand/WC19_Docs/TransportChecklist.html
RESNA's Position on Wheelchairs Used as Seats in Motor Vehicles	http://resna.org/resources/position-papers/RESNAPositiononWheelchairsUsedasSeatsinMotorVehicles.pdf
BC Motor Vehicle Act Motor Vehicle Restraint Systems and Booster Seats Safety Regulations (SOR/2010-90) Child Seating and Restraint Systems	http://www.bclaws.ca/EPLibraries/bclaws_new/document/LOC/freeside/--%20M%20--/46_Motor%20Vehicle%20Act%20RSBC%201996%20c.%20318/05_Regulations/29_26_58%20-%20Motor%20Vehicle%20Act%20Regulations/26_58_11.xml#part_division36
Sunny Hill Health Centre for Children	http://www.bcchildrens.ca/Services/SunnyHillHealthCtr/default.htm
The Family Fund	http://givinginaction.ca/giahome.htm

Through the Looking Glass	http://www.lookingglass.org/index.php
Translink: Access Transit Secretariat	http://www.translink.ca/en/Rider-guide/Accessible-Transit/Access-Transit?Access-Transit-overview.aspx
WestJet: Guests with special needs	http://www.westjet.com/guest/en/travel/special-arrangements/special-needs/index.shtml

Ministry of Children and Family Development

At Home Program	http://www.mcf.gov.bc.ca/at_home/
At Home Program Guide	http://www.mcf.gov.bc.ca/at_home/pdf/ahp_guide.pdf
17-Year-Old Disability Assistance Applicants	http://www.hsd.gov.bc.ca/factsheets/2005/17yr_old_PWD.htm
At Home Program Medical Benefits Transition to Disability Assistance - July 2013	http://www.mhr.gov.bc.ca/clientinfo/docs/At-hometrans.pdf
Health Supports for Children in Care and Youth Agreements	http://www.mcf.gov.bc.ca/foster/pdf/health_supports_cic.pdf

Ministry of Social Development and Social Innovation

Medical Equipment and Devices - Overview	http://www.gov.bc.ca/meia/online_resource/health_supplements_and_programs/mem obility/index.html
Medical Equipment and Devices - Policy	http://www.gov.bc.ca/meia/online_resource/health_supplements_and_programs/mem obility/policy.html
Medical Equipment and Devices - Procedures	http://www.gov.bc.ca/meia/online_resource/health_supplements_and_programs/mem obility/procedures.html
Medical Equipment and Devices - Authorities and Responsibilities	http://www.gov.bc.ca/meia/online_resource/health_supplements_and_programs/mem obility/authorities.html
Medical Equipment and Devices - Forms and Letters	http://www.gov.bc.ca/meia/online_resource/health_supplements_and_programs/mem obility/forms.html
Ministry Office Directory	http://www.hsd.gov.bc.ca/contacts/city.htm

BC Government

Personal Supports - Home	http://www.personalsupports.bc.ca/psp/supports/equipment_and_assistive_devices.page?keyword=Equipment+and+Assistive+Devices~
Employment and Assistance for Persons with Disabilities	http://www.eia.gov.bc.ca/pwd/eapwd.htm
Employment Program of British Columbia (EPBC)	http://www.gov.bc.ca/meia/online_resource/employment_programs_and_community_services/epbc/
Equipment and Assistive Technology Initiative (EATI)	http://www.bcpsn.org/