

Addressing Life Long Care Needs in Duchenne Muscular Dystrophy *Every Single One - PPMD*

Physical Therapy
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Acknowledgements

PPMD

Continually supports education locally, throughout communities, nationally, and internationally.

We appreciate all that they do to advance our understanding of this disease and bring families, professionals, and researchers together as a team!

Contributing Therapists to ESO's

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* Laura Case * Leslie Vogel*

Stage 1: • At Diagnosis

Stage 2: • Early Ambulatory

Stage 3: • Late Ambulatory

Stage 4: • Early Non-ambulatory

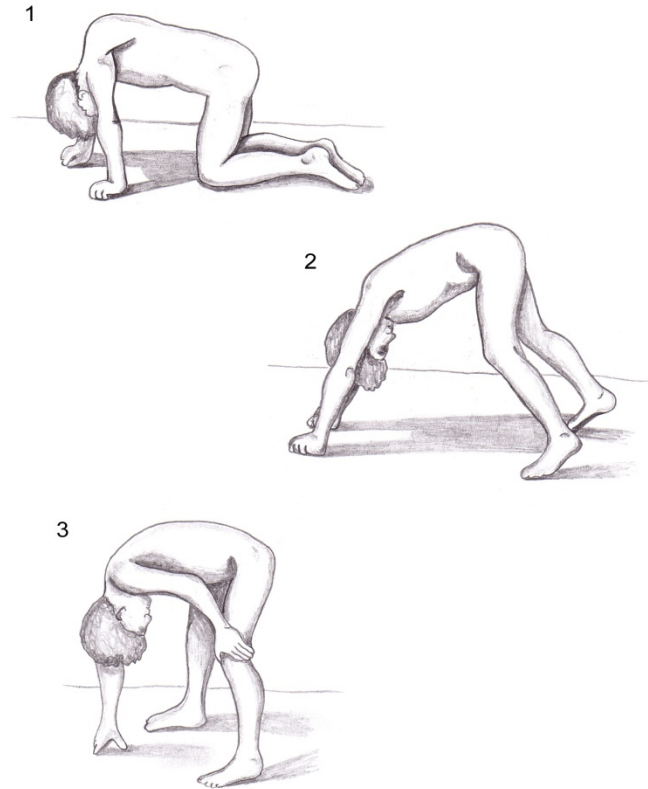
Stage 5: • Late Non-ambulatory

At Diagnosis

- May appear clumsy, may fall more frequently than peers
- May have motor and/or language delays
- May have difficulty keeping up with peers
- Initiating steroids

Early Ambulatory (Elementary School)

- Common compensatory movements appear
 - (+) Gower's sign
 - Waddling gait/Duchenne jog
 - Possible toe walking
 - Can climb stairs though may use compensatory pattern
- May have difficulty keeping up with peers



Late Ambulatory (Late Elementary into Middle School)

- Increasingly labored gait
- Loss of ability to climb stairs and rise from floor independently
- Increasing lower extremity contractures as ambulation declines
- Increasing reports of back pain
- Weight gain

Late Ambulatory (Late Elementary into Middle School) (cont.)

- Limited community mobility
- Increased fatigue
- Increasing difficulty with activities may lead to decreased motivation to participate
 - May further contribute to decline
 - Disuse atrophy
 - Muscle wasting when the muscle is no longer as active

Early Non-Ambulatory (Middle School into High School)

- Typically able to sit independently, though bed mobility may become more difficult
- Difficulties with activities of daily living (ADLs)
 - bathing, toileting, and transfers
- Increased risk of developing scoliosis
- Increased upper extremity tightness and weakness
 - difficulty with handwriting
 - difficulty with self feeding
- Initiation of BiPAP

Late Non-Ambulatory (High School and Beyond)

- Limited upper extremity function, postural maintenance, and bed mobility
 - Increasing need for caregiver assistance for all ADLs and repositioning
- Increasing use of respiratory support
- Increasing reports of chronic pain
- Further decline in community participation and engagement

Positive Things to *Do and Encourage*

Contributors to Muscle Tightness and Joint Contractures

- Weakness
- Prolonged positioning
- Muscle imbalance
 - If can't move through a full movement actively then likely to develop tightness
- Fibrotic changes in muscle

Stretching

- Stretching has not been shown to improve, maintain or prevent loss of range of motion
- However, potential benefits of brief stretching (i.e. 30 seconds) may include:
 - Temporary improvement in blood flow to muscle
 - Temporary decrease in pain
 - Temporary feeling of well-being
 - Temporary increase in tolerance to stretch
 - Temporary decrease in muscle stiffness
 - Continued movement through joint's full available range of motion

Stretching - Areas of Tightness

- Legs
 - Calf muscles: tightness limits ability to move ankles up
 - Hamstrings: tightness limits ability to straighten leg
 - Hip muscles: tightness limits ability to lie flat on stomach or move leg towards middle of body
- Arms
 - Elbow flexors: tightness limits ability to straighten elbow
 - Forearm pronators: tightness limits ability to turn hand palm up
 - Long finger flexors: tightness limits ability to straighten fingers
 - Shoulder extensors: tightness limits ability to bring arms overhead

Stretching – Areas of Tightness

- Natural history data is indicating that tightness develops in calf muscles early on
- Tightness in knees and hips begins to emerge ~1 year prior to loss of ambulation when spending more time in sitting

Positioning

- Avoid long periods of time in one position that facilitate known areas of tightness
- Frequent position changes are recommended
- However, alternative positioning can be used for prolonged stretch

Orthotics – Night Splints

- Resting brace ankle foot orthotic (AFO) not to be used with walking
- Starting at young age to improve tolerance and wear time
- Most useful before tightness develops
- Continue to prevent further contracture development
- Custom-made braces with wrap around forefoot design
- Set at comfortable end range
- (Scott et al 1981, Hyde et al 2000)



<http://cascadedrafo.com/products/rafo-3.5-softy>



Photo courtesy of Ted Ryder,
Hangar Prosthetics and Orthotics

Orthotics (cont.)

- Goal 8 hours/night
- If not tolerated
 - Try alternating each side at night
 - Use for daytime positioning when seated or lying
- May transition to daytime wear when non-ambulatory for ease of movement in bed at night

Orthotics – Wrist Hand

- Wrist hand orthotic (WHO) with extension under the fingers for tight long finger flexors
- Goal is to keep fingers straight
 - Set at comfortable end range of wrist and finger extension



<https://www.benik.com/adults/wrist/volar-pan-extension>

Other Positioning Devices

- Nada Chair kiddy up may assist with long sitting for hamstrings
- Knee immobilizers for night time wear, with night splints, may be considered



www.nadachair.com



<https://www.medicaldepartmentstore.com/FLA-Orthopedics-Universal-Knee-Immobilizer-p/fl37914.htm>

Optimized Positioning - Seated

- Straight upright posture
- Feet supported
- Knees in line with feet and hips – not apart
- Laptop /computer screens at eye level



Seated Positional Changes/Prolonged Stretching

- Arms hanging down at sides to stretch elbow
- Legs propped up with toes pointed up to stretch knees
- Use of full recline with leg extension in power wheelchair to stretch hips and knees



Optimized Positioning - Lying

- On back
 - Legs as straight as possible
 - Pillow under knees can relieve pressure on the back
 - Support tight muscles with pillow/blanket/towel
 - Limit time with knees positioned apart
- On side
 - Support the body with pillows/blanket/towel
 - Support between thighs can reduce strain on the back and hips

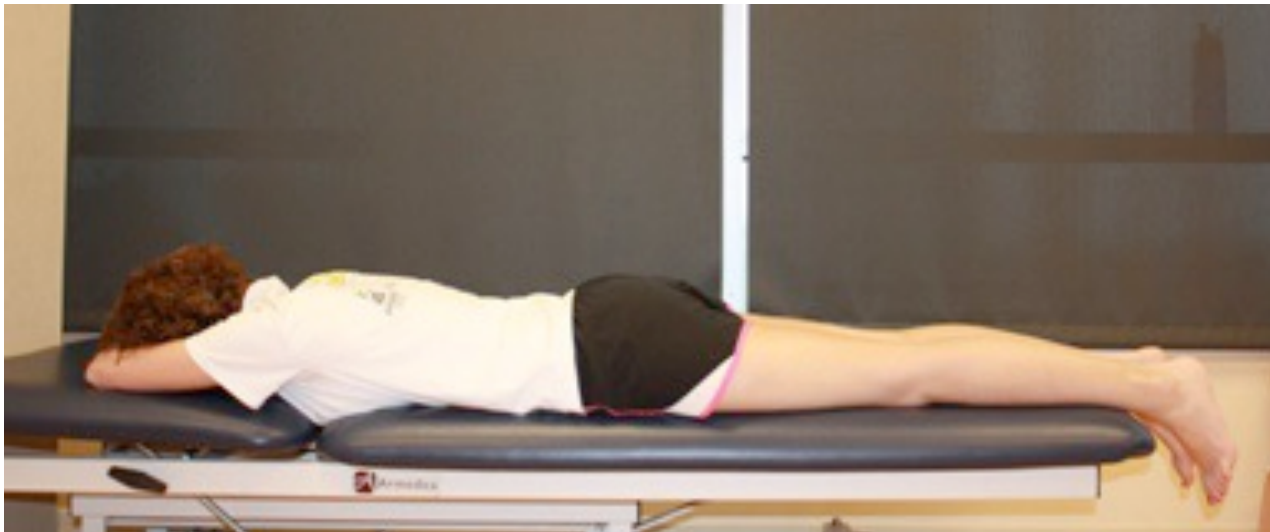
Lying Positional Changes/Prolonged Stretching

- Lying on back
 - Arms overhead to stretch shoulders
 - May need support under knees to reduce strain on low back
 - Legs as straight as possible to stretch hips and knees
 - Keep legs close together



Lying Positional Changes/Prolonged Stretching

- Stomach lying
 - Feet off the bed to stretch hips and knees if tolerated
 - May need support under pelvis for tolerance if hips are very tight



Standing Positional Changes/Prolonged Stretching

- This can be accomplished through using of standing devices
- After loss of functional ambulation

Benefits of Continued Standing

- Leg range of motion (Paleg, 2013; Townsend, 2016)
- Bone mineral density (Paleg, 2013)
- Lung function (Galasko, 1995)
 - reduced risk for scoliosis (Galasko, 1995)
- Bowel function (Paleg, 2013; RESNA position paper on standing)
- Psychosocial

When to Initiate Supported Standing

- In late ambulatory or early non-ambulatory stages
 - When standing and walking in good alignment becomes difficult (Case, 2018)
- Prior to development of significant hip, knee, and ankle tightness
- Delay can limit tolerance, comfort, and adherence

Standing Program

- Minimum of 5 days per week
- 60 to 90 minutes
 - Can be broken into smaller time increments
- Gradual increase in duration
- As upright as tolerable

Static versus Dynamic Standers

- Both allow gradual transition from seated to standing position
- Tray for functional work surface
- Chest strap for increased trunk support and to feel secure



<https://easystand.com/product/evolv-medium/>



<https://permobilus.com/products/childrens-corpus-vs/>



Dynamic Standers on Wheelchair

- Advantages
 - Can move in and out of standing without additional transfer
 - Theoretically can transition into standing independently
 - Can drive in standing
 - May provide additional benefits for maintaining bone density (RESNA; Ward 2004)
- Disadvantages
 - Standing feature often denied so can delay time to obtain power wheelchair
 - Footplates not as angle adjustable to accommodate heelcord tightness
 - Device trying to accomplish two things so may not fit as well for both

Static Sit to Stand Standers

- Advantages
 - Footplates are angle adjustable to accommodate heelcord tightness
 - Can be obtained at any time without impacting attainment of power wheelchair
 - Can attain a more fully upright position
- Disadvantages
 - Must be transferred into the device
 - Cannot typically move independently from sitting to standing
 - Not mobile in the device

Exercise

What we know now!

Exercise: What do we know?

- No exercise leads to muscle atrophy
- Too much exercise increases muscle breakdown (rhabdomyolysis)
 - Muscle pain up to 24-48 hours after over activity
 - Myoglobinuria (coca-cola colored urine)
- Eccentric (lengthening) contractions are more damaging to the muscle cell
 - Going down stairs
 - Jumping on trampoline
- Boys with DMD are 40% less active than age matched peers (McDonald, 2000)

Exercise Recommendations

- Energy conservation involves doing activities
 - With the least amount of effort
 - In the most efficient way
- Pacing
 - Plan ahead for “big days” to allow rest periods
 - Use assistive devices for long distances to save energy for activities important for your child
 - Have your child rest before he gets tired

Exercise Recommendations

- Age appropriate recreational activities as opposed to strengthening
- Activities should be fun and promote self esteem and social interaction
- Stay submaximal, avoid overexertion

Exercise: What do we know?

- “No use is disuse study”:
 - In late ambulatory and early non-ambulatory phases submaximal assisted bicycle training was safe and feasible and demonstrated stable motor functioning over 24 week period (Jansen, 2013)
- In late ambulatory phase active/active-assisted/assisted arm bicycle training vs active range of motion
 - Both groups improved to varying levels (Alemdaroglu, 2015)

Exercise Recommendations (cont.)

- Cycling
 - Assisted cycling or cycling without excessive resistance
 - Avoid hills or give assistance
 - Add power when needed
 - Training wheels or three-wheeled bike to help prevent falls



Exercise Recommendations (cont.)

- Aquatics/swimming
 - Easier to move in the water
 - Buoyancy of water supports body weight
 - Allows freedom of movement especially in late ambulatory and non-ambulatory phases
 - Develops independence and confidence
 - Provides opportunity for life long recreational activity



Outpatient Therapist's Role

- Monitor your child's range of motion (flexibility)
- Assist in development of a stretching, positioning, and activity program that meets your child's needs
- Assist in obtainment of orthotics, equipment, and assistive devices/technology
- Periodic/consultative model of care with the exception of
 - Early development
 - Times of transition
 - Post fracture/post surgery

School Therapist's Role

- Promote safety, positioning, and independence within the school setting to enhance learning

Equipment *Across the Lifespan*

Goals of Equipment

- To enhance independence and function
- To reduce the impact of the disease
- To increase safety and prevent injury
- To optimize quality of life

Risk of Loss of Ambulation

	Best predictor of loss of ambulation within 1 year	Reduced risk of losing/unlikely to lose ambulation within 1 year	Best predictor of loss of ambulation within 2 years	Reduced risk of losing/unlikely to lose ambulation within 2 years
NSAA	9/34 (Ricotti, 2016)		NSAA 13/34 (Ricotti, 2016)	18/34 (Mazzone, 2013) 16/34 (Ricotti, 2016)
10m	>10-12 sec (Birnkrant, 2018)			<= 7 sec (Mazzone, 2013)
Gowers (supine to stand)	Inability to stand from supine/>30 seconds (40% lost ambulation McDonald, 2013; 57% lost ambulation Mazzone, 2016)	Ability to rise from the floor (2.3% lost ambulation McDonald, 2013)		<=10 sec (Mazzone, 2013)
6MWD	<325m (40% lost ambulation McDonald, 2013)			>= 330m (Mazzone, 2013)

Special Considerations

- Equipment needs discussed regularly and requested proactively.
- Select a piece of equipment that will be appropriate now, but also for the next 3-5 years.

Process for Obtaining Equipment

- Obtain referral
- Establish equipment team
- Complete equipment evaluation
 - Opportunity to discuss, see and try equipment
- Consider home evaluation for ADL equipment or when space is a concern
- Obtain quote
- Write letter of medical necessity
- Prepare for denials and need to appeal

Who is Involved?

- Need for team approach (Mannlein, 2008)
 - In more rural areas this may mean pursuing equipment through a wheelchair or seating clinic at a larger institution
 - Child and Family
 - Referring physician
 - Therapist/clinician
 - Equipment provider/vendor/supplier

Mobility Equipment

When is Mobility Equipment Needed

- Walking becomes tiring or difficult
- Frequent falls
- When family activities are being limited
- (Case, 2018)

Adapted Strollers

- Advantages

- Compact
- Easy to use
- Easy to transport
- Relatively low cost
- Non medical appearance

- Disadvantages

- Child has to be pushed
- Limited growth and positioning
- Best on smooth, flat surfaces
- Appearance for an older child may be “babyish”

Adapted Stroller (cont.)



<https://www.adaptivemall.com/mainbug.html>

<https://www.convaid.com/product/cruiser-5/>

Transport Wheelchairs

- Advantages
 - Fold for easy transport
 - Possibility of back-up to power chair
- Disadvantages
 - Child has to be pushed
 - Limited growth and positioning
 - Best on smooth, flat surfaces



https://www.amazon.com/Medline-Lightweight-Transport-Wheelchair-Handbrakes/dp/B007WA1ZG4/ref=sr_1_1_sspa?ie=UTF8&qid=1536691050&sr=8-1-spons&keywords=transport+wheelchair&psc=1

Ultra-lightweight Manual Wheelchairs

- Advantages
 - Customizable seating
 - Easily transported
 - Can be grown
 - May be able to push self for short level distances
 - Possibility of back-up to power chair
 - Less costly than power wheelchair
- Disadvantages
 - May need to be pushed for longer distances
 - Limits of ability to serve as back-up to power chair depending on size

Ultra-lightweight Manual Wheelchairs Features

- Maximal growth
- Capability to add power assist components
- Height adjustable push handles
- Solid seat and back
- Pressure relieving washable seat cushion
- Thigh guides/hip adductors
- Swing-away footrests
- Lateral supports
- Seat belt
- Chest strap if needed for transport
- Angle adjustable footplates
- Airless tire inserts
- Headrest if needed for transport
- Transport package



<https://permobilus.com/product/aero-x/#>

Power Assist Wheelchairs



Push-rim activated (alber e-motion, Quickie Xtender, alber twion)



Permobil SmartDrive



Rio Mobility Firefly

<http://www.alber-usa.com/downloads/prospekte.html>

<http://www.max-mobility.com/smartdrive/#mx2pluspushtracker>

<https://riomobility.com/firefly/>

Power Assist Wheelchairs

(cont.)



- Advantages

- Easily transportable
- Decreased effort required to push
- Increases independence with mobility

- Disadvantages

- Cost
- Increased weight
- Require charging
- Less maneuverability than a joystick drive power wheelchair

Scooters

- Advantages
 - Independent mobility
 - Relatively inexpensive
 - Can be broken down for transport, but cumbersome
- Disadvantages
 - Poor postural support
 - Decreased stability, especially 3-wheeled models
 - Large turning radius
 - Less maneuverability than a joystick drive power wheelchair
 - Child must transfer out for transportation in vehicle

Scooter Considerations

- Lower seat height or add support under feet
- Back support pillow
- Bring handlebars closer to child
- Add seat belt
- Four wheeled for increased stability
- Watch for hip abduction
- Should be considered a mobility device alone, rather than a mobility and seating device



<https://www.pridemobility.com/c/go-go-travel-mobility>

Power Wheelchairs on Folding Frames

- alber e-fix



e-fix®

<http://www.alber-usa.com/produkte-rollstuhl-zusatzantrieb/zusatzantriebe-fuer-rollstuehle/rollstuhlzusatzantrieb-efix.html>

- EZ Lite Cruiser



<https://www.ezlitecruiser.com/products/ez-lite-cruiser-standard-model>

Power Wheelchairs on Folding Frames (cont.)

- Advantages
 - Fold for easy transport
 - Customizable seating
- Disadvantages compared with traditional joystick drive power wheelchair
 - Less maneuverability
 - Not as durable
 - Lack of power seating features limiting
 - independence/ease with transfers and activities of daily living
 - positioning options

Power Wheelchairs



- Advantages
 - Independent mobility
 - Power seating features to increase
 - comfort
 - independence with transfers and activities of daily living
 - Can be used on varied outdoor surfaces
 - Customizable
- Disadvantages
 - Expensive
 - Requires means of transport and accessible environments

Power Wheelchairs Drive Configurations

- Front Wheel Drive
 - Improved curb and obstacle climbing capabilities
 - Better in soft terrain (grass/gravel)
 - Provides tight maneuvering
- Mid-Wheel Drive
 - Smallest turning radius/increased maneuverability
 - Beneficial for indoor use
- Rear Wheel Drive
 - Heavy outdoor use or high speeds
 - Large turning radius
 - Limited curb climbing ability
- (Mannlein, 2008)

Power Wheelchairs

Recommended Features

- Pressure relieving washable seat cushion
- Thigh guides/hip adductors
- Lateral supports/custom-molded seat back
- Seat belt
- Chest strap if needed for transport
- Angle adjustable footplates
- Airless tire inserts
- Headrest if needed for transport
- Transport package
- Power tilt
- Power recline
- Power seat elevator
- Power leg elevation
- Power anterior tilt
- Power standing
- Tray
- Swing away/retractable joystick
- **Case 2018 includes nice justifications**



<https://permobilus.com/products/power-wheelchairs-by-permobil/front-wheel-drive/>

Power Wheelchairs

Recommended Features

- With disease progression
 - Alternative drive controls
 - Blue tooth capability
 - Microlite switches
 - Ability to carry ventilator
 - Elbow supports
 - Gel overlays
 - Mount for smartphone/tablet

Equipment to Support Activities of Daily Living

Bathroom Equipment

- Safety is a significant concern due to risk for falls with wet surfaces and balance requirement for many bathing and toileting tasks
- Home health OT or PT eval prior to purchase to ensure it will be able to be utilized in the home

Bathroom Equipment (cont.)



- Initially minimal equipment and modifications
 - Grab bars
 - Raised toilet seat
 - Simple bath or transfer bench
 - Hand held shower nozzle

Bathroom Equipment (cont.)



- As physical function declines
 - Ideally would have accessible bathroom with roll-in shower
 - Can be difficult to obtain funding for bathing and toileting equipment so long term needs should be considered when equipment is selected

Bathing and Showering

- Rolling chair with tilt-in-space option and commode cut out
- Roll-in Shower
 - Aquatec Ocean VIP
 - Shower Buddy Roll in Buddy and Tilt
 - Raz Attendant Tilt



<http://myshowerbuddy.com/roll-in-buddy-with-tilt/>

Bathing and Showering

(cont.)

- Stander tub or shower
 - Similar equipment with transfer attachment
 - Columbia Omni Bath Shower Commode Transfer System
 - Shower Buddy Tub Buddy with Tilt



<https://www.healthproductsforyou.com/p-columbia-omni-reclining-bath-shower-and-commode-transfer-system.html>

Toileting

- Combined shower/commode chair for bowel management
- Hand held urinal for bladder management
 - Slide forward on seat or use power recline feature
 - Uri-bag: more flexible and portable urinal
- Consider tear away or snap athletic pants for clothing management

Transfer Equipment



- NIOSH guidelines recommend lifts be used when weight exceeds 35lbs (Waters, 2007)
 - Decreasing risk of injury to both child and caregiver

Transfer Equipment: Sliding Boards

- When core and arm strength are good
- When performing level or downhill transfers
- When space limits use of sling system
- Standard or sliding seat/disc (i.e. Beasy Board)



<http://www.beasyboards.com/>

Transfer Equipment: Sling Systems

- Stand alone mobile devices (i.e. Hoyer lift)
 - Manual (hydraulic) or electric
 - Can be moved from room to room
 - Portable versions available
 - Can be used to transfer patient on/off floor
- Wide adjustable wheeled base for stability but difficult to use in bathrooms and confined spaces



<https://www.spinlife.com/Hoyer-Advance-H-Patient-Lift-Quick-Ship-Manual-Patient-Lift/spec.cfm?productID=81056>

Transfer Equipment: Sling Systems (cont.)



- Permanent overhead lift systems (i.e. Sure Hands/Voyager)
 - Typically within a bedroom or to adjoining bathroom
 - May not be covered by insurance

Transfer Equipment: Sling Systems

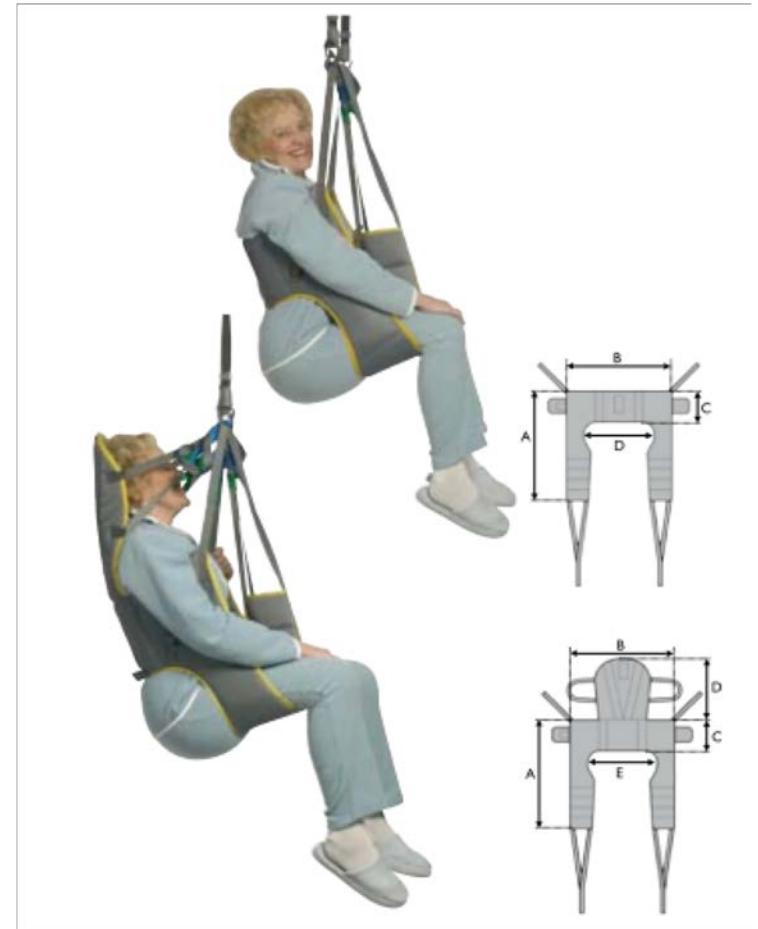
- U shaped padded sling with head support
 - Can be put on/taken off while in a seated position
 - Does require significant degree of trunk, hip and knee flexion



https://www.amazon.com/Drive-Medical-Padded-Support-Medium/dp/B002VWK140/ref=sr_1_3_a_it?ie=UTF8&qid=1538418891&sr=8-3&keywords=hoyer%2Bsling&th=1

Transfer Equipment: Sling Systems (cont.)

- Dress toileting sling
 - Large opening allows easy access for adjustment of clothing and toilet transfers



<http://www.oflynnmedical.com/product/invacare-dress-toileting-sling/>

Hospital Beds

- Semi electric hospital beds with standard mattresses typically covered by insurance
 - Elevation of head and foot of bed for comfort/transitions to sitting
 - Railings provide surface to pull on for assist with turning
 - Hand crank to raise and lower bed
- Fully electric beds and higher end pressure relieving mattresses can be more difficult to get funded

Assistive Technology

Assistive Technology: Why to Consider?



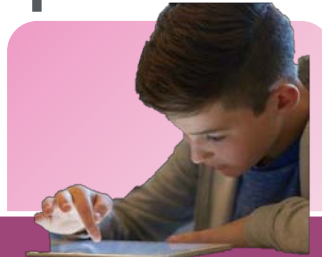
- More focus on using assistive devices to facilitate arm function and encourage participation in social activities to improve quality of life (Lue, 2017)
- High priorities/highly valued activities (Bergsma, 2016)
 - Eating and drinking independently
 - Preparing food
 - Personal hygiene
 - Computer use
 - Touching their face
 - Brushing their teeth
 - Toileting
 - Driving their wheelchair
 - Repositioning at night
 - Shifting while seated

Low, Middle and High Technology Options



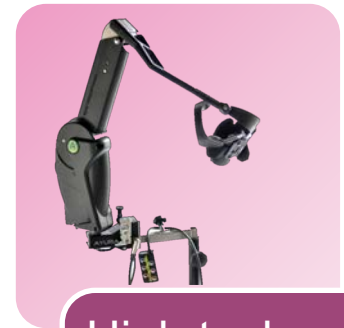
No/low tech

- Twist-n-write pencil
- Pencil grips
- Modified utensils



Mid tech

Environmental controls
Smart Home systems
Computer/iPad/Chrome books
Speech-to-text software
Text-to-speech software
Word prediction software
Read and Write app from Chrome store
Kami app from Chrome store
Siri on iPad
Google type with speech built into tools
Sonocent note taking software



High tech

- Mouse modules on power w/c
- Mobile arm supports

Dynamic Arm Supports



WREX: Wilmington
Robotic Exoskeleton

<http://jaecoorthopedic.com/products/WREX%3A-Wilmington-Robotic-EXoskeleton-Arm.html>



Armon Ayura

<http://qartal.org/wp-content/uploads/2016/04/arm-on-ayura-2.jpg>



JACO robotic arm

<https://www.abilities.com/community/buzz/64b-jaco.html>

Thank You!

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