

Get Smart with Spinal Muscular Atrophy

Nicole Wilkins
Occupational Therapist
Sunny Hill Health Centre for Children
Vancouver, BC, Canada



Objectives

- Identify one potential solution for seated postural support with a client with SMA
- Identify one potential solution for power mobility setup with a client with SMA
- Become more familiar with current level evidence
- Identify one method for controlling the mouse or an electronic aide to daily living through the power wheelchair

Flip Back Custom Armrests

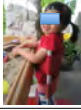


Custom Seating System



Maliyah

- SMA 2
- 18 months in video, 3 years in photos
- Ranger power wheelchair with power stand up & tilt feature
- Fanthera manual wheelchair
- Flexi-stand stander
- Hips located, no scoliosis



- Fanthera manual wheelchair (sublight)
- Short indoor distances, flat firm surfaces
- Isidra commercial cushion
- Padded upholstery back
- Horizontal chest strap
- Pelvic belt



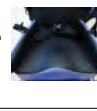
Spinal Muscular Atrophy

- Genetic Disease motor (and sensory – Mentis, 2013) nerve cells in the spinal cord. Progressive weakness torso then distal muscles, affects sitting, walking, crawling, breathing, swallowing.
- SMA Type 1: most severe form, 1st yr. not sit, rapid deterioration weakness, life expectancy 2-3 yrs (ls not roll or sit, life expect < 30 mo, lb – live longer)

- SMA Type 2: symptoms 6-18 mo, become weaker, seated support, scoliosis by 4 yrs, not walk. Life expectancy young adult
- SMA Type 3: milder weakness, child or teen yrs, walk into adulthood, lifespan can be normal, (lba – mild with motor impairment, walk up 3 yrs, lllb – mild with normal motor dev until 3 yrs)
- SMA Type 4: milder, adult onset weakness


Foam and Plywood Seat Cushion

- 1 1/2" soft foam, 4 way stretch fabric
- full length pelvic laterals
- mild ischial block with weighted seat
- custom built-in pommel for hip abduction
- 1" AES padded seatbelt



Foam and Plywood Backrest

- Sacral block
- Swing-away curved trunk laterals, symmetrical
- Dynaform neoprene shoulder straps
- Whitmyer headrest
- Padded footrest




- Ranger power wheelchair
- Power tilt
- Stand-up feature
- Commercial seat cushion
- curved backrest
- Protect deep contour curved backrest
- Dynaform shoulder/chest strap, pelvic belt
- Midline mount compact joystick





Hannah

- SMA Type 2
- 3 years old
- Invacare TDX Spree power wheelchair (elevator, power tilt)
- Drives with central MEC joystick
- Custom seating system
- No scoliosis, lateral X femoral heads uncovered



ASL Micro-Extremity Joystick



Visual Display & Controls

- Mounted on left armrest
- Uses left hand to directly touch the visual display to change drive speeds, and use power tilt and elevation (via joystick)



Future Directions - Hannah


- May need more sensitive switches closer to joystick
- Over-ride tilt to allow her to drive in more tilt (head balance)
- Obtain soft neck collar for transport or bumpy/hilly terrain, stroller
- Trial a more occipital headrest
- Over time, may need fuller length, more flexible trunk supports or a trunk orthotic (abdomen open)

Future Directions - Maliyah


- Difficult to provide curved trunk lateral supports at the correct height on trunk with stand up chairs as they move higher as do armrests
- Difficult to provide contoured or thick seats or backrests as need keep pelvis close to pivot point
- Maybe obtain trunk orthotic (open abdomen) later

Aidan

- SMA Type 1b
- 7 years old
- Invacare TDX Spree power wheelchair (elevator, power tilt)
- Drives with central MEC joystick
- Custom seating system
- ventilator & g-tube



Custom Seating System



Foam and Plywood Seat Cushion

- 1 1/2" soft foam, 4 way stretch fabric
- pelvic laterals & built-in pommel
- mild ischial block with weighted seat
- 1" ischial obliquity support (right side high)
- 1" AES padded seatbelt




Future Directions - Aidan

- Obtained prescription trunk laterals molded by Orthotist (Dr. refused TLSO)
- Need to interface orthotic side supports to seating trunk lateral components
- Need to measure respiratory function with trunk orthotics, seated postural control measures, client comfort, pressure map
- Obtain 2 in 1 switch cord for power/mode function

Chase

- SMA Type 1b, 10 years old
- Invacare Ranger Trique SP power wheelchair (power tilt)
- Drives with MEC joystick (both hands)
- Custom seating system
- Spinal rod instrumentation T1 to lumbosacral junction 2007 (B6-Regene T2-L3)
- Bilateral hip dislocation




Arm Support

- Dynamic elbow/upper arm cuffs (joystick/switch use, iPad and iPod use)




Elevating Legrest

- Elevating legrest (manual handle to adjust)
- No footrests (feet/ankles fully contracted & sensitive)




Foam & Plywood Backrest

- Scoliosis (right side forward, convex left, concave right)
- Swing-away curved trunk laterals (right side high)
- Whitmyer occipital headrest
- Padded armrests & footrest
- Dynaform neoprene shoulder straps




Custom Seating System

- Foam in Box custom contoured backrest
- Foam and plywood memory foam seat with ROHO insert
- Swing-away trunk laterals
- Whitmyer headrest and left lateral support



Foam and Box Backrest (memory foam "squared")



ASL Micro-Extremity Joystick & custom sensitive switches



Chase Computer Use


- Uses MEC joystick to control laptop mouse & Smartboard
- Uses iPad, iPod using direct finger touch
- Upper arms are braced in midline using dynamic loc-line mount with upper arm cuffs
- Elbows are braced together in midline against front edge of his trunk laterals

Aidan x-rays



Aidan Computer



- Uses Mac touchpad bluetooth to laptop (positioned low right side to allow wrist flexion)



Foam and Plywood Seat (ROHO insert, built-in pommel)



Whitmyer Headrest





Future Directions - Chase

- More flexible trunk lateral supports (milder support, more movement), then but soft
- Power footrest that can go up (hip flexion), or out
- 2 in 1 switch to control power & mode
- Obtain more EADES

Kalin

- 15 years old, SMA 2
- Quantum 6000, custom T joystick, remote plus (older electronics)
- Spinal rod instrumentation
- Has 2 gel seat cushions, foam in box backrest
- Has lateral & posterior tilt




Kalin Seating System



Kalin Computer

Mouse through joystick on power wheelchair

Mouse through built-in touchpad




Future Directions

- Decrease # switches but increase functions (3 switches total for power, mode, left click)
- Obtain an ACM box so can integrate power tilt and lateral tilt into joystick (eliminate 2 switches)
- Obtain drag/drop, right click software (dragger), abbreviation/expansion software

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Considerations for Power Mobility

- power mobility at a young age (18 months)
- power tilt (more degrees for driving)
- Stand-up power for those with adequate trunk, head control
- sensitive joysticks (often midline mounted)
- mounting joystick lower than forearm support
- using sensitive finger switches close together
- Over time integrate seating from switches into controller (i.e. joystick)


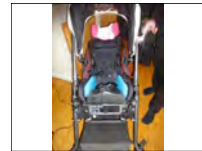
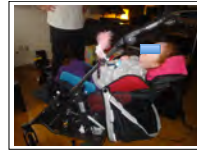
Computer & EADL Use

- Use the most efficient and direct computer access method (often built-in touchpad on laptop)
- if very weak, computer access through power wc controller (sensitive joystick, sensitive switches for left/right clicks) – not all power wc electronics are created equal
- Incorporate EADL's into power wc for easier access, or obtain separate devices

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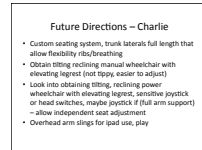
Charlie-Anne

- 3 years old
- SMA Type 1b
- Stroller with custom seat & back
- Reclined to manage secretions, breathing, needs frequent ++ suctioning
- Vocalizes, some words

Charlie-Anne Computer

- Has a Tobii C12 EyeGaze Computer for speech output, learning, play

Future Directions – Charlie

- Custom seating system, trunk laterals full length that allow flexibility (bc/breathing)
- Obtain tilting reclining manual wheelchair with elevating legrest (not top, easier to adjust)
- Look into obtaining tilting, reclining power wheelchair with elevating legrest, sensitive joystick or head switches, maybe joystick if full arm support – allow independent seat adjustment
- Overhead arm slings for quad use, play

Literature – main findings

- Trunk orthotics DO NOT stop the progression of the spinal curve (1,7)
- Most articles suggest that trunk orthotics are used prior to (not after) spinal instrumentation for postural support, all balance BUT ensure aware respiratory function and allow for abdominal freedom or be non-rigid (1-7)
- Rigid full trunk orthotics can impair respiratory function (1-4,5,7)
- Power mobility young (20 months) in SMA 1 & 2 (1,8)

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4. Fujik A, Andohgawa M, Park H. Prevalent SMA. *J Orthop Phys Ther* 2007;14(5): 213-212
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7. McKinnon MS, Blain PA, Oakley M et al. A Dutch guideline for the treatment of scoliosis in neuromuscular disorders. *Scandinavian* 2008; 1:4
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What do we need to work on?

- More research & use of measurement tools
- Share experiences, consensus on seating practices
- Work together as a team (seating, orthotics, respiratory)
- Develop new products (flexible thin trunk lateral supports)

Use More Measurement Tools

- Respiratory: (Bipap, ventilator, oximeter, chest measurement, observation rib/abdomen/chest movement)
- Seating Posture: (Seated Postural Control Measure – SPCM, goniometry, sitting height)
- Use of client outcome rating scales (Canadian Occupational Performance Measure – COPM)
- Pressure mapping
- Hand/Arm function
- Pain Scales

Considerations for Positioning

New style trunk laterals!

- Flexibility (respiratory function) vs support (posture)
- Thin (arm/hand function)
- Integrate orthotic laterals into seating system?

Alternate Style trunk orthotics!

- Semi rigid, open abdomen trunk Orthotics (pre-spinal instrumentation)?

- Pelvic laterals past knee & pommel to abduct hips
- Soft seat and backrests (lung expansion, comfort – air or gel after rod instrumentation)
- Recline for SMA 1
- Padded occipital style headrests, soft neck collar transport
- Elbow/forearm support, elbow blocks, flexible arm support/fit assist supports
- Soft padding on footrests, dynamic footrests (up/out)

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