What is SMA?

- Loss of function of spinal motor neurons causing progressive weakness of muscles
- Muscles include extremities, respiratory muscles
- Sensation is normal
- Brain function is normal
What is the lower motor neuron?

MOTOR UNIT = Anterior Horn Cell + Nerve Fibre + Muscle Fibre

In SMA some of the Anterior Horn Cells are affected and fail to provide the link between the Brain and the Nerve Fibres.
Therapeutic Intervention

- Physical therapy
- Occupational therapy
- Bracing/ Orthotics
PT

- Gross motor development
- Maintain flexibility, ROM and strength of the LE’s and minimize contractures
- Upright weightbearing
- Mobility/ Ambulation
OT

- UE/hand function
- Maintain flexibility, ROM and strength of the UE’s/hands and minimize contractures
- Perceptual motor development and experiences
- Social emotional growth and development
Child’s occupation

- Importance of play
- Importance of therapeutic activities
Coordinate with other disciplines

“Best care”

Important to coordinate all care with others managing child’s care (therapists, doctors, teachers, caretakers)
Therapy Intervention

- Respiratory
- Maintain flexibility and avoid contractures
- Facilitate sensorimotor development and exploration
- Innovations for play and positioning
- Strengthening/ Exercise/ Fitness
- Energy conservation
- Mobility
Therapy Intervention

- Maximize a child’s function
  - A player and a learner
- Promote and facilitate independence in functional activities
- Sensorimotor experiences
Developmental therapy

- Maximize function
- Optimize play
- Facilitate mobility
Developmental Handling

- To promote movement
- Facilitate skill development
- Exploration of environment
- Movement control
- Movement experience and comfort with movement
OT/PT - Developmental handling
Therapy (OT, PT)
Promote and facilitate independence in functional activities

- Expectations for independence
- Self care activities
- Interaction within the community
- School
- Adaptive equipment
- Positioning
Occupational Therapy
Maximize flexibility, ROM, strength of UE and hands and avoid contractures

- Maintain strength and ROM of trunk, UE’s and hands as able
- Assess functional abilities as related to strength
- Develop compensatory strategies that maximize the child’s physical abilities
- Watch for fatigue and overuse, endurance
- Splinting for UE’s
Facilitate sensori-motor development and exploration

- Play
  - Body awareness, vestibular and tactile input and experiences
- Developmental postures and activities
- Adaptive positioning and innovations for play
- Toys
  - Types and controls
- Environmental accessibility
  - Environmental controls/ computers
Sensorimotor experiences

- Movement experiences
- Body awareness
- Motor planning
- Motor control
Sensorimotor development and exploration
Social-emotional growth and development

- Behavior
- Motivation
UE/ Hand function

- Fine motor development
- Bilateral coordination
- Shoulder girdle integrity
- Splinting
- Positioning
Hands and splints
Slings and springs
Play and positioning
Play and exploration
Perceptual motor development and experiences

- Importance
- Perceptual motor development
Adaptive equipment

- Facilitate independence
- Play
- School
- Resources
Play and adaptations
Adaptive toys
Adaptive switches/ toys
Switches for environmental control
Assistive technology

- Evaluation
- Resources
Physical Therapy
Respiratory

- Respiratory play
- Postural drainage
- Breathing exercises
- Positioning for improved respiration
  - TLSO, Benek’s vest, Abdominal binder
Respiratory
Maintain mobility and avoid contractures

- Prevent deformity
  - Range of motion exercises
  - Stretching to preserve or increase flexibility
  - Positioning
  - Splinting
- Infancy throughout lifespan
- Be Pro-active
Passive stretching?

Florence, Glanzman, Krosschell, Maczulski, Main, Morrison 2006, Wang's Journal of SMA Care
Gross Motor Development

- Stability
- Mobility
- Skill acquisition
- Compensatory strategies
Strengthening/ Exercise/ Fitness

- Maintain strength as able
- Assess functional abilities as related to strength
- Train to use to maximal advantage
- Watch for fatigue and overuse
  - Strengthening does not change progression
  - Too much can cause overfatigue
Maintain flexibility
Strengthening, exercise and fitness
Energy conservation

- Do not overwork areas of increased weakness—may lead to overuse syndromes, pain and increased weakness
- Scheduling to optimize energy and strength
BOB GETS OUT THE TAPE MEASURE AFTER ANOTHER DISAPPOINTING WINTER RIDE:

WOULD YOU SAY MY QUADS ARE MIGHTY?

MIGHTY PUNY.
Upright weightbearing

- **Physiologic advantages**
  - Prevention of contractures
  - Decrease osteoporosis
  - Prevention of UTI’s
  - Increase cardiopulmonary tolerance
  - Development of head and trunk control
  - Strengthening
Upright weightbearing

- Quality of life enhanced
- Increased self esteem
- Participation in group activities with greater independence
- Positioning for upper extremity use
Upright weightbearing

- Positioning/ Alignment
- A-frame/ Parapodium/ Supine/Prone standers
  - A start towards ambulation
- Adaptations for control and alignment in standing
- Critical time periods
  - 12-24 months
  - Post surgery
Monitor equipment needs

- Braces and splints
- Assistive devices
  - Walkers and crutches
- Standers/ Upright Positioners
- Wheeled mobility
  - Strollers
  - Wheelchairs/ Scooters
  - Stand and Go’s
Ambulation

- Pros and cons
- When and how
- Training and practice
- Effects of strength
- Changes with TLSO
Bracing for standing and ambulation

- AFO’s
- KAFO’s
- Ischial weightbearing KAFO’s or LLB’s
- HKAFO’s
- RGO
- Parapodium
- Adaptations to aid in ease of donning and doffing
Braces for standing and ambulation
Bracing and assistive devices for standing/ambulation
Learning to walk with KAFO’s
Gait training
**Stand and Deliver: A Guide to Self-Propelled Standers**

By Ginny Paleg, PT and Steve Mauricio, PT

### Standers at a Glance

<table>
<thead>
<tr>
<th></th>
<th>Standers</th>
<th>Action Wheelchairs</th>
<th>Wheelchairs by Locomotion</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Product</strong></td>
<td>Standing</td>
<td>Action Wheelchairs</td>
<td>Wheelchairs by Locomotion</td>
</tr>
<tr>
<td><strong>Strengths (+)</strong></td>
<td>- paint on all frames, push handle, seat, seat adjust independently of one another and lab all directions</td>
<td>- great seat comfort, non-slip seat for greater and greater strength</td>
<td>- can escape in and out of wheelchair, facilitate use of extra seat, adjust forward or backward can be customized to accommodate even the tallest people</td>
</tr>
<tr>
<td><strong>Weaknesses (-)</strong></td>
<td>may not be wide enough for kids in space needed</td>
<td>need an unnecessary to adjust</td>
<td>- simple use only</td>
</tr>
<tr>
<td><strong>Does the person need to be left side and right side?</strong></td>
<td>no</td>
<td>no</td>
<td>yes</td>
</tr>
<tr>
<td><strong>Easily adjustable for children users?</strong></td>
<td>no</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td><strong>Does it have a safety belt for a car?</strong></td>
<td>no</td>
<td>yes</td>
<td>no</td>
</tr>
<tr>
<td><strong>Cost without accessories</strong></td>
<td>$1,925</td>
<td>$2,795</td>
<td>$3,490 to $4,895</td>
</tr>
<tr>
<td><strong>Wheel size</strong></td>
<td>16&quot;-19&quot;</td>
<td>16&quot;-19&quot;</td>
<td>16&quot;-19&quot;</td>
</tr>
<tr>
<td><strong>Chair size range</strong></td>
<td>9&quot;-28&quot;</td>
<td>9&quot;-28&quot;</td>
<td>9&quot;-28&quot;</td>
</tr>
<tr>
<td><strong>Wheelbarrow</strong></td>
<td>no</td>
<td>yes</td>
<td>yes</td>
</tr>
<tr>
<td><strong>Different items available</strong></td>
<td>no</td>
<td>yes</td>
<td>yes</td>
</tr>
<tr>
<td><strong>Comes with tray</strong></td>
<td>yes</td>
<td>no</td>
<td>yes</td>
</tr>
<tr>
<td><strong>Optional height adjustments</strong></td>
<td>yes</td>
<td>yes</td>
<td>yes</td>
</tr>
<tr>
<td><strong>Lateral support adjusted independently</strong></td>
<td>yes</td>
<td>yes</td>
<td>yes</td>
</tr>
<tr>
<td><strong>Child can access brakes</strong></td>
<td>no</td>
<td>yes</td>
<td>yes</td>
</tr>
<tr>
<td><strong>Has high back and head support</strong></td>
<td>yes</td>
<td>yes</td>
<td>yes</td>
</tr>
<tr>
<td><strong>Arms for standing</strong></td>
<td>yes</td>
<td>yes</td>
<td>yes</td>
</tr>
<tr>
<td><strong>Control and must support</strong></td>
<td>yes</td>
<td>yes</td>
<td>yes</td>
</tr>
<tr>
<td><strong>Telephone number</strong></td>
<td>(800) 210-7694</td>
<td>(800) 223-4896</td>
<td>(850) 774-0531</td>
</tr>
</tbody>
</table>

This chart is meant as a guide for families and professionals to assist them in determining if a self-propelled stander is appropriate and which model offers the options that they need. Equipment changes all the time, so please check with your equipment supplier to make sure that the information below is still accurate.

---

**Easy Stand Mover and Easy Stand 500 with Mobile Option**

- http://www.easystand.com
- Can be tilted 100 degrees to allow optimal positioning.
- Mobile stander is comfortable.
- Wheels can be tilted to allow optimal positioning.
- Can be used independently of one another, lab all directions.
- Can be used in and out of wheelchair.
- Multiple seat pad locations can be used as a child and stander.
- Need an additional wrench to some adjustments and need good upper extremity strength to propel.
- Mobile stander may be too short for many users.
- Wheels can be tilted for a child with short torsos or below average height. Tools needed to make adjustments.
- Pans are vary high off the ground.

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---

**Alder**

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---

**Wheelchair**

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**Standers**

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Mobile standers
Standers
Adaptive equipment at home
Bathing and toileting
Car seats
Lifts and ramps
Wheeled mobility

- Positioning
  - When to correct alignment and when to accommodate alignment
- Adaptive seating
- Carriers for medical equipment
- Safe mobility and transportation
Wheelchairs

- Manual or power
- Options
  - Elevating legrests, swing away legrests
  - Adjustable height arms
  - Seating and support
  - Headrest
  - Tilt in space and/or recline systems
  - Power controls - placement and types
  - Seat belt or harness
  - Tray - to assist in ADL and for UE support
Wheelchairs and strollers
Recreational intervention

- WC sports
- Hippotherapy
- Adaptive swimming
- Modified bowling
- Summer camp programs
- FUN- of utmost importance
Windsurfing
Hippotherapy
Skiing
Canoeing at camp
Companion canines
Thank you!!