



## Families of SMA

*Research. Support. Hope.*

### Empowering Families of SMA-- Navigating your Child's Course of Therapy for Best Outcomes

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### Empowering Families of SMA-- Navigating your Child's Course of Therapy for Best Outcomes





Empowering Families of SMA--  
Navigating your Child's Course of Therapy  
for Best Outcomes

Objectives include discussion of:

- Role PT, OT, Speech and Family
- Therapy and Adaptive Equipment
- Movement, Strength and Standing
- Minimizing Contractures and Pain
- Optimizing Oral-Motor and Written Communication Skills



Empowering Families of SMA--  
Navigating your Child's Course of Therapy  
for Best Outcomes

Consensus Statement for Standard of  
Care in SMA

1. Confirm SMA diagnosis
2. Manage breathing
3. Manage eating and nutrition
4. Manage movement and daily activities
5. Prepare for illness

Journal of Child Neurology, Volume 22  
Number 8, August 2007, 1027-1049



## Physical Therapy

**PT's Role** is to maximize active movement / strength, prevent / minimize contractures / pain in the trunk and lower extremities.

Achieved by:

- Range of Motion
- Positioning
- Strengthening
- Equipment Recommendations
- Bracing/Splinting



## Occupational Therapy

**OT's role** is to improve ability to participate in meaningful occupations (activities),

Achieved by:

- Maximizing movement / strength and preventing / minimizing contractures / pain in the upper extremities
- Improving access to age-appropriate environments, (eg: toys, writing/drawing activities, computers, etc.) at home and school
- Optimizing independence / safety while eating, grooming, dressing, bathing, toileting, etc. "ADL's"



## Speech Language Therapy

**Speech Therapist's Role** is to optimize communication, social interaction, and feeding/swallowing abilities:

- Types of Communication: Speech, Augmentative Communication, Sign Language, Gestures
- May benefit from amplification, ideas for increasing breath support/positioning, voice output device and/or picture boards



## OT, PT or ST??????

- Feeding and Swallowing may be addressed by Speech, OT, or both disciplines
- Augmentative Communication needs are typically evaluated by both Speech and OT and PT may involved with positioning
- Respiratory therapy may be done by PT
- Younger the child-often more the overlap



## Role of Family / Role of Therapist

.Expectation for two-way communication

.Best outcomes:

- consideration of values / lifestyles of both child and family
- Collaborative home programs
- Incorporation of recommendations into daily routine

.Focus on function

.Start prevention early



## Role of Family / Role of Therapist

*Buzz* Terms for Intervention and Frequency

“Evidence-Based Practice”:

- using research evidence for clinical decision making
- Pediatric therapists have many barriers to this process
- Research in Pediatrics often have low strength levels



## Role of Family / Role of Therapist

### *“Frequency”*

- Intensive: 3-11 x a week
- Weekly/Bimonthly: 1-2 x week or every other week
- Periodic: monthly or less often at regularly scheduled intervals
- Consultative: episodic or as needed



• Bailes AF et al, 2008



## Role of Family / Role of Therapist

### How do WE decide?

#### Key Issues to keep in mind:

- Functional goals
- Rate of progress
- Amount of clinical decision making needed
- “Critical” periods
- Priorities
- Episodes of Care
- Insurance



Ustad et al, 2009



## Adaptive Equipment

- What is available for Children?
  - Lots!!!
- What are the issues with children and equipment?
  - Funding
  - Growth
  - Support vs Independence
  - Process to get equip



## Adaptive Equipment

When does my child need equipment?

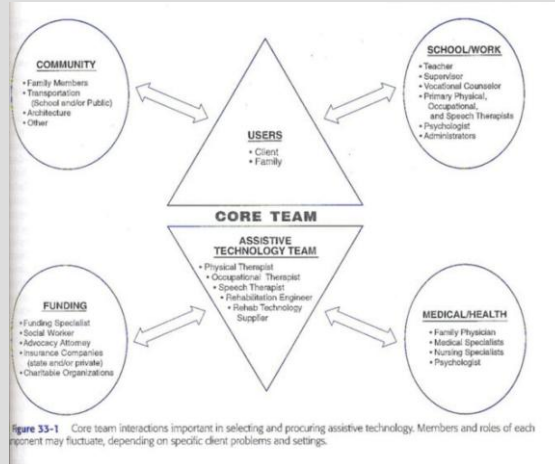
- Mobility: school, community, home
  - Strollers, wheelchairs-power vs manual
- Positioning: Sitting, standing, side lying
  - Adapted chairs, standers
  - ADL's
  - Special Needs Car seats / Vests
- Recreation/Exercise
  - Adapted tricycles
- *May need to try equipment first in environment it will be used—ask vendor or therapist if this is an option*





# Adaptive Equipment

## • Who Decides????



# Adaptive Equipment

## • Who Pays?

- Insurances
  - Check policies-they don't cover everything
- Family Support
- Community Options Programs
- The family

## • Loan Closets

[www.fsma.org](http://www.fsma.org)  
 (Support->Community->Daily Life->Equipment Pool)  
 1-800-886-1762

## • Insurance

- Keep it for 5 years
- May pay after 3 years if outgrowing
- Manual w/c before power
- May cover car seats
- May cover bath equipment
- May cover standers







## Adaptive Equipment

When choosing consider:

- Amount and type of support needed.
- Where the equipment will be used:
  - Adult turning radius: RWD=33"  
MWD=23"  
FWD=25"
- How much space is there for storage?
- Who will be using/user friendliness?
- Goals of the equipment?
- The child's growth and equipment's ability to grow



## Adaptive Equipment

*Why is Equipment Abandoned?*

1. Lack of Consideration
2. Ease of Procurement
3. Performance
4. Priorities

❖ *Nearly 30% abandoned*



Phillips B et al, 1993



## Power Mobility

*What skills must a child have to use power mobility?*

- Cognitive Level of at least 2 yrs. of age
- Understand Motoric Language
- Understand Spatial Concept
- Functional Skills to Operate w/c

Campbell SK et al, 2006; Jones MA et al, 2003



## Adaptive Bathroom Equipment Considerations

### Child/Family:

- Head and/or trunk support?
- Recline or Tilt in Space?
- For toileting or just bathing?
- Physical ability of caregivers?
- Other family use bathroom?



### Environment:

- Dimensions?
- Width of doorway?
- Tub versus roll-in shower?





## Adaptive Bathroom Equipment Options

### Inflatable Bed Bath



### Tub Slider/ Glider



### Shower/Commode



### Roll-in Shower

### Mobile Lift

### Ceiling Lift



## Safe Travel

Restraint must meet or exceed applicable  
Federal Motor Vehicle Safety Standards  
(FMVSS 213)

Questions? Locate a Child Passenger  
Safety Technician in your area

[www.safekids.org](http://www.safekids.org)

Safe Kids Near You -> Find Coalitions and  
Events Near You -> Enter State



## Safe Travel

Consider size, postural-control, respiratory status, needs of family, and vehicle

Stay rear-facing as long as possible

- Some convertibles RF up to 32-35 pounds
- Graco My Ride (< 40 pounds)

May use rolled towels/blankets along side of head/body, but not underneath

Plan ahead for Wheelchair-Accessible Van

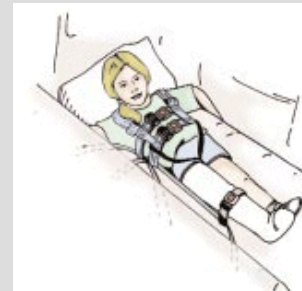


## Safe Travel--Type 1

Respiratory Tolerance Test

Cosco Dream Ride  
Car Bed (<20 #, <26 inches)

Modified EZ on Vest  
(>12 months; >20 #; <48 inches)





## Safe Travel--Type 2

Five-point harness restraint

Lateral head support

High shoulder harness settings

Examples:

- Britax Traveller Plus
- Special Tomato
- Roosevelt (Velcro cap)



## Safe Travel--Type 3

Five-point harness restraint

High shoulder harness settings

Examples:

- Ride Safer Vest
- Radian (Sunshine Kids)





## Safe Travel- Wheelchairs

ANSI RESNA WC/19: voluntary standard developed that addresses issues of using wheelchairs as a seat in a bus or van

In general, wheelchairs not designed for transportation

If possible, use Car seat

ANSI (American National Standards Institute and Rehabilitation)

RESNA (Rehabilitation Engineering and Assistive Technology Society of North America)



## Safe Travel- Wheelchairs

If unable to transfer, need:

- Four clearly marked crash-tested attachment points on the frame for tie downs
- No slack on straps
- Recline angle=within 30 degrees of upright



Wheelchairs that meet WC19:  
<http://www.rercwts.org/WC19.html>





## Is my child able to get stronger?

“Functional ability declines as the body mass of the individual overcomes the limited motor neurons.”

(Krosschell K, McGovern D, Maczulski J PT, OT, and Orthotic Considerations with SMA  
APTA Combined Sections Meeting 2009)

### Exercise:

- Exercise activates molecular and cellular cascades that support CNS function, plasticity and protection against damage

(Grandard 2005; Biondi 2008; Charbonnier 2007)



## Is my child able to get stronger?

- General Guidelines:
  - Promote movement
  - Incorporate into daily activities
  - Modify position for most benefit
  - Strengthen for functional abilities
  - Avoid disuse!!!



(Grandard 2005; Biondi 2008; Charbonnier 2007)





## So how do we maintain my child's strength?

### Use of gravity-eliminated planes for active movement



Mobile  
Arm  
Supports



## So how do we maintain my child's strength?

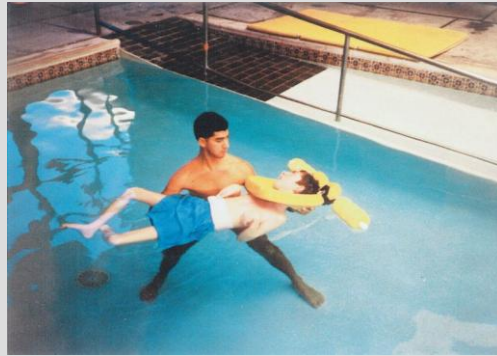
Suspension for the child's arm or leg will provide opportunities to move without the resistance of gravity.

- Arm anchor (pivot):
  - above the shoulder
  - forearm with the thumb up
- Leg Anchor (pivot):
  - above the hips
  - toes are pointing up





So how do we maintain  
my child's strength?



### Aquatherapy

Cunha et al 1996



So how do we maintain  
my child's strength?



Functional Use  
during Daily Activities



Play in Weight-bearing  
Positions (Prone)



## Would e-stim help my child get stronger?

### TES vs FES

- Evaluated effect of low-intensity night-time therapeutic electrical stimulation (TES) on arm strength / function in SMA Type 2
- Randomized controlled trial with a 6-month baseline control period



## Would e-stim help my child to get stronger?

### Results:

- No statistically significant differences:
  - Strength
- No significant change:
  - excitable muscle mass
  - function on the Pediatric Evaluation of Disability Inventory (self-care domain)

- Fehlings, et, al , 2002



## Standing

- Maintaining the ability to stand and/or walk for as long as possible.
  - Improve lung function compared to those who only use a wheelchair
  - Scoliosis deteriorates more slowly  
(Robinson D et al, 1995)
- Case Studies that report prolonged ambulation with bracing  
(Evans GA et al, 1981; Tangsrud SE et al, 2001)
- Critical Periods:
  - 12-24 months of age
  - Post surgery



## Standing

- Other Benefits:
- Peer Interaction
  - Increased self esteem
  - Positioning for upper extremity use
  - Prevention of contractures
  - **Decrease in osteoporosis**
  - Prevention of UTI's
  - **Increase in cardiopulmonary tolerance**
  - Strengthening for head and trunk control
  - Enhance digestion





## Standing

### Braces for Standing:

- SMO's
- AFO's
- KAFO's
- Ischial Wt Bearing KAFO's
- HKAFO's
- RGO
- **Watch weight of assistive devices**



## Standing

### Devices for Standing:

- Walkers
- Gait Trainers
- Parapodium
- Static Standers
- Mobile Standers





## Why are children with SMA at risk for contractures and pain?

Hypotonia (decreased resistance to passive stretch)

Joint Laxity: may lead to subluxation/dislocation

Growth/ weight gain: places a greater demand on available muscle strength

Weakness

- more proximally than distally
- more in legs than in arms
- affecting some muscles more than others



## How do we minimize contractures and pain?

Muscle Imbalance / Common Resting Position:

- Adducted shoulders (arms at side)
- Pronated (palms down) forearms
- Ulnar deviation (wrists bent towards little finger)
- Flexed (bent) knees
- Plantarflexed (toes pointed down) ankles





## How do we minimize contractures and pain?

- Maintain passive range of motion (caregiver moving)
- Optimize positioning
- Bracing/ Splinting
- Maintain active range of motion (child moving)
- Maintain strength



## How do we minimize contractures and pain?

### Benefits of Passive Range of Motion / Massage

- Maintains skin elasticity, muscle length, and joint integrity
- Improves circulation (blood and lymphatic)
- Provides sensory input



### Possible Consequences of Lack of ROM:

- May affect skin integrity
- May cause pain due to contractures / hypersensitivity
- May decrease function







## POSITIONING



For Type 1, positioning options are limited by respiratory status

Important to work toward or maintain tolerance to upright positioning



ROM and positioning work best when part of daily routine

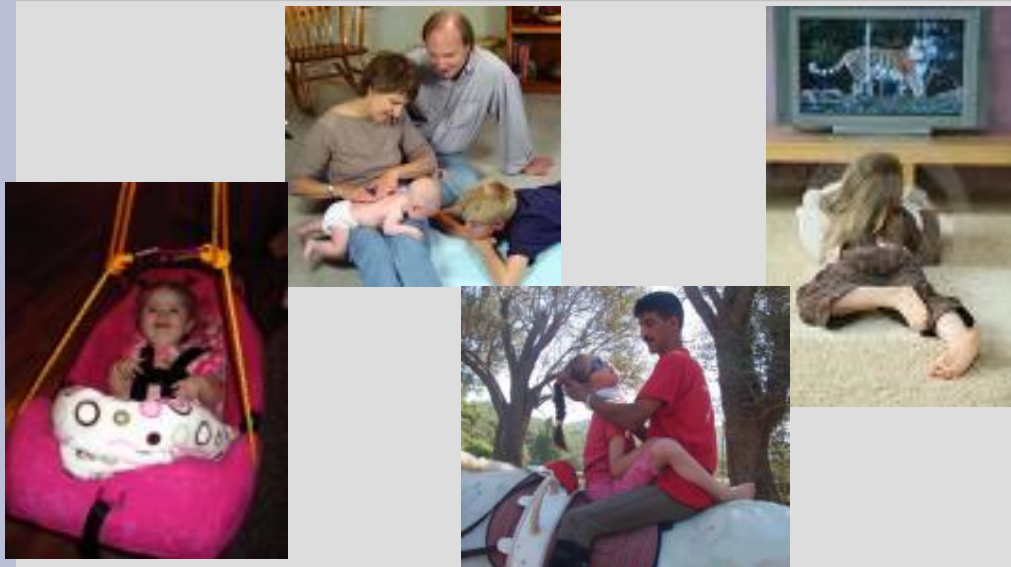


## POSITIONING





## POSITIONING



## Braces and Splints

Thoracic Lumbar  
Sacral Orthoses  
(TLSO)





## Braces and Splints

- Recommend TLSO with abdominal cut-out to minimize compromise in oxygen saturation  
(Frownfelter, Cook, Massery, Bernardoni 2006)
- However, be sure to monitor respiratory effects
- Custom Fabrication by Orthotist using light weight thermoplastic material
- Posterior or bivalved opening
- Ventilation Holes



## External Supports (Less Rigid)



Benik Trunk Support  
Theratogs



Kinesiotape



## Braces and Splints

Goals of night splints/braces: prevent contractures

Provide low-load prolonged stretch (prolonged ROM) for gradual tissue lengthening

- Night Ankle-Foot Orthoses
- Knee Immobilizers
- Resting Hand Splints
- Elbow Extension Splints
- Dry Rice or Dry Beans in Sock



## Braces and Splints

Goals of day splints/braces: alignment for function

Examples:

- Ankle foot orthosis
- Wrist Supports

Weight considerations important





## Optimizing Oral-Motor Skills

Regarding ROM and Strength:  
Let's not forget the facial muscles ☺

- 15 individuals with SMA; age 6 to 21 years of age
- Jaw muscle strength 56% of normal
- UE strength 35-40% of normal (Koch 1992)
- Less efficient muscles and fatigued 30% faster
- Weakness and reduced effort may alter craniofacial development and produce malocclusions

Granger et al 1999



## Optimizing Oral-Motor Skills

Range of Motion of Jaw reduced (1/2 range of vertical opening and protrusion)

Parents report children:

- take longer
- have difficulty with fibrous / hard foods

Oral aversion and hypersensitivity (to tastes, textures, and smells) is common after:

- medical procedures involving the airway
- limited exposure to food







## Optimizing Oral-Motor Skills

What can we do to help with jaw mobility and sensory hypersensitivity?



For Infants / Toddlers:

- Offer pacifier (esp during tube feedings)
- Help to orally explore a variety of safe textures

For School-Age Children

- Active jaw movement with a mirror
- Open/close and forward/back
- If approved by physician, offer gum in an upright position



## Optimizing Oral-Motor Skills

- Provide positive stimulation to gradually desensitize
- Gently massage the jaw, inside and out
- May use small battery-powered Mini or fingertip massager
- Try battery-operated toothbrush
- Use of a chewy tube or theratubing
- Involve in social aspect of family meals
- Be respectful of child's response
- Pediatric dentist





## Optimizing Pre-Academic Skills

Research shows individuals with SMA usually have average or better intelligence.

Van Gontard A et al 2002; Sigford BT et al 1998; Riviere J 2002

Therefore, very important to consider adaptations for play to facilitate development of cognitive-perceptual abilities.

(Cook 2002; Berry 2003; Desch 1986)



## Optimizing Written Communication Skills

Federal Law (PL 94.142) Individuals with Disabilities Act (1997): All students are entitled to a free and appropriate public education.

Written communication is an integral component in academic success.

In most states, at least one Assistive Technology Specialist (for consultation).

Assessment in actual environment important.







## Adaptive Writing Techniques versus Using Computers?

Team decision (child, parent, teacher, school OT and Assistive Technology specialist)

If writing is a goal, may benefit from:

- alternating hands
- slant board/easel
- adaptive writing utensils
- modified assignments



Voice-dictation and/or word-completion software are options



## Computer Access Options

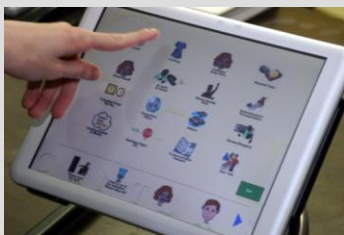
Eye Gaze

Voice Activated

Touch Screen

Switch

Trackball





## If using switch, many options:



Micro light



Mercury



Jelly Bean



Wobble



## Role of Family / Role of Therapist

.Expectation for two-way communication

.Best outcomes:

- consideration of values / lifestyles of both child and family
- Collaborative home programs
- Incorporation of recommendations into daily routine

.Focus on function

.Start prevention early



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Thank you to all of the children  
in our pictures and in our hearts.

