

**The Role of Therapy in your
child's life.**

Being a part of the team!

2018 Annual CureSMA Conference



Make today a breakthrough.

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Disclosure

Karen Patterson, PT, MS, PCS

- Medical Advisory Committee for CURE SMA
- Clinical Investigator for AVXS –101 – CL – 303
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Plan for Today...

Part 1: Therapy basics-what you need to know

- How to be an active member of your child's therapy team
- What we know and don't know regarding therapy interventions
- How often /How much?
- Exercise/activity and play ideas
- Different models of therapy
- Other types of therapies and up and coming and technology

Plan for Today...

Part 2: The Changing Landscape of SMA

- What does this mean for therapy?

Part 3: Tips for Caregiver

- Body mechanics for lifting and transfers

Time for Discussion

Part 1

Therapy Basics



Don't Overlook
the Basics!
The Basics
are the Best!

Families' Frustration

- Lack of experience with SMA by therapist.
 - You may not be able to choose your therapist.
- Uncertainty about quantity needed for your child.
 - Limited availability, frequency or times
- We do not have much evidence to support which “type” of therapy is best.
- Delays in getting therapy, treatments....
- You are pioneers... but that can be exhausting!!!!



Start building your team



- **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 (need goals!)
 - Recognize landscape is changing
 - Start prevention early!

The Therapy TEAM



You and your child are the captains!

- Occupational Therapist (OT)
 - Adaptive aids, fine motor/hand skills, feeding, equipment
- Physical Therapists (PT)
 - Strength, stretch, sit, walk, posture, balance, equipment
- Speech Therapist (SP or SLP)
 - Language, jaw issues, feeding
- Augmentative and Alternative Communication (AAC)
 - All forms of communication, aided or unaided (could be OT/PT/ST)

Other Team Members

- **Physiatrist (Rehab Medicine Physician)**
 - Physician that oversees rehab care
- **Orthopedist**
 - Physician for bones and muscles
- **DME (durable medical equipment) vendor**
 - Wheelchairs, strollers, walkers, standers, special chairs, car seats, beds
- **Orthotist**
 - Braces (AFOs, SMOs, TLSOs, KAFOs)
- **Others**
 - Massage, swimming, horseback riding, chiropractor, acupuncture

Interventions and Frequency

- What do we know and what don't we know?
- “Evidence-Based Practice”:
 - Using research evidence for clinical decision making
 - Pediatric therapists have many barriers to this process
 - Research in Pediatrics often has low strength levels



Revised Consensus Statement for Standards of Care in SMA (2016):

North America

- Jacqueline Montes
- Kristin Krosschell
- Allan Glanzman
- Richard Gee
- Leslie Nelson

Europe

- Marion Main
- Elena Mazzone
- Anna-Karin Kroksmark
- Carole Vuillerot
- Caron Coleman
- Agnieszka Stepień

APAC

- Kristy Rose

Strengthening and Exercise



What we KNOW

Consequences of muscle weakness:

- Creates challenges with movement and function = MOVE LESS
- Impacts breathing, being upright and putting weight through legs
- May limit positioning and play
- Can lead to muscle tightness and asymmetries that could lead to joint contractures, pain, falls

What we THINK we know

Therapeutic Exercise and Activity

- May be of value to maintain and increase strength for function.
 - But how much strength?
- We can often predict which muscles will get tight and which muscles will be the weakest.
 - But we cannot always be sure what activities and exercises will best address these issues.
- Maintaining and/or increasing range of motion (ROM) is beneficial
 - But does it improve function?

What we KNOW: Exercise

- Daily exercise is safe in ambulatory SMA and should be encouraged. (Montes et al, 2015)
 - May need to exercise for longer period of time (year?) to see benefits
 - Warrants further study
- Strengthening Exercise in a home-based, individualized program may maintain strength and increase sense of well-being for children and adolescents with type 2 and type 3 SMA. (Lewelt et al, 2014)
 - Well tolerated and may increase strength in children and adults with SMA, but further study in larger groups are necessary.
- Aerobic Cycle exercise improves VO_{2max} in those with SMA 3
 - Without causing muscle damage, but it also induces significant fatigue. (Madsen et al 2014)
- **Important to use the muscle you have!!!**
 - muscle that is not used gets weaker
- **We do not yet know when or if exercise leads to overwork induced muscle damage.**

Consensus Statement on Exercise

Strengthening:

- Muscles with antigravity strength – can use some resistance
- Neck muscles – resistance NOT recommended
- Muscle less than antigravity – eccentric not recommended

Aerobic Exercise:

- At least 30 minutes , 2-3x/week (minimum), 3-5x/week (optimal)
- Encourage various forms

Flexibility



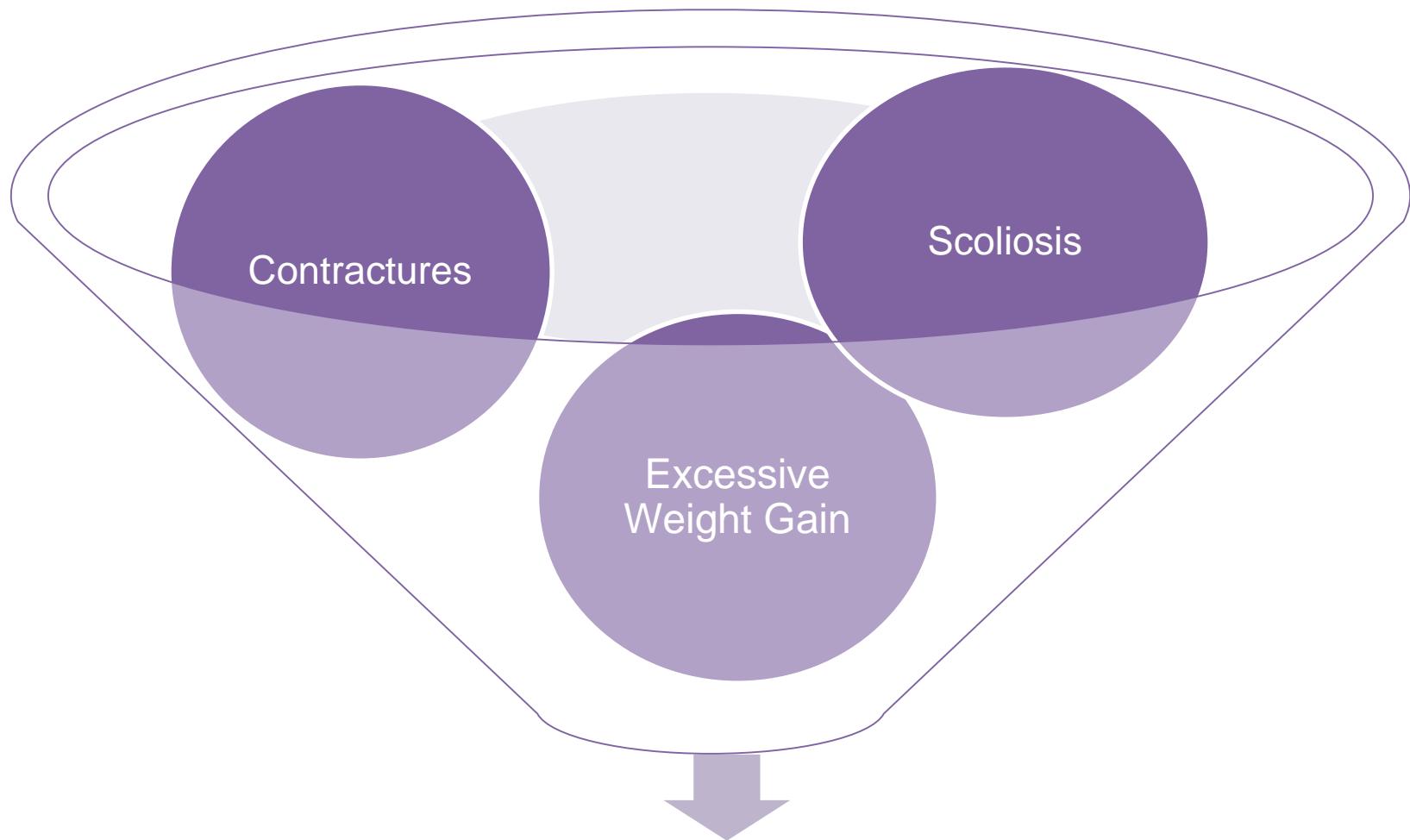
Flexibility: What we KNOW

Why are children with SMA at risk for tightness?

- Weakness :
 - Difficulty moving in/out of positions and remain in some positions longer than others
- Imbalance:
 - affecting some muscles more than others
- Growth/ weight gain:
 - places a greater demand on available muscle strength



Flexibility: What we KNOW



Loss of Function in Type 2 & 3

Flexibility: What we KNOW

Benefits of Range of Motion (stretch), Movement and Massage

- Function and Positioning
 - More options for function, play, activities of daily living
 - Potentially enhances stability
- Pain and Comfort
 - A tight joint can hurt
- Dressing and hygiene can become difficult
- Improves circulation (blood and lymphatic)
- Provides sensory input

How often/How long: What we THINK we know

- Holding a stretch longer is better for other conditions
 - Splints or braces may hold positioning longer
- Consistency is better:
 - Daily routines that are established will assure that stretches are done more frequently.
- Prioritize Muscles:
 - Your therapist can help you do this

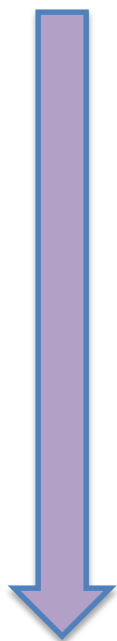


Consensus Statement: Stretching

- May be accomplished using bracing, standers, active assisted stretching
- Duration:
 - To IMPROVE length: >60 minutes (standers and bracing)
 - To MAINTAIN length: hold at end range for at least 60 sec.
- Frequency:
 - Bracing – use both day and night
 - Optimally= 5 – 7x/week, Minimal = 3 – 5x/week
- Standers:
 - Duration: up to 60 minutes
 - Optimally = 7x/week, minimal 3-5x/week

Therapeutic Intervention Recommendations

More
Important



Less
Important

Non Sitters	Sitters	Walkers
Stretching/ Range of Motion	Stretching/Range of Motion	Exercise
Seating Systems/Postural and Positioning Supports	Wheelchairs	Transfers and Mobility Training
Play/Toys	Play/Toys	Stretching/ROM
Education	Bracing	Aquatic Therapy
Assistive Technology/Adaptive Equipment	Seating Systems/Postural and Positioning Support	Balance Exercises
Bracing	Standers	Bracing
Wheelchairs	Transfer and Mobility Training	Fatigue Management
Chest PT	Mobile Arm Supports	Activities of Daily Living (ADL) Management
Transfer and Mobility Training	Aquatic Therapy	Ambulation Devices
Mobile Arm Supports		
Exercise		
Aquatic Therapy		

Positioning for Function

- On your back (supine)
- On your stomach (prone)
- Sidelying
- Sitting
- Standing/Walking

Handouts for activity ideas

- What are good activities you can do in all positions?
- Ideas for building a PVC sling support
- Standing with some support
- Aquatic therapy and some head supports
 - Otteroo Neck Float <https://otteroo.com/>
 - Waterway babies neck ring
<http://www.waterwaybabies.com/contact-us-order/>
 - <http://www.theraquadatics.com/aquafit.html>

General Considerations

- Respiratory Status
- Work toward increasing tolerance or maintaining upright, as possible
- Joint contractures

Supine (on your back)

- Often well tolerated
- Consider the use of wedges to start working towards upright – monitor respiration
- Small pieces of foam cut into different shapes, and weighted bean bags to support and position extremities
- Promote midline



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Sitting

- Vary amount of incline and upright
- Optimal amount of support to promote function (varies)



Special
Tomato
Activity Chair



Leckey
Squiggle
Activity Chair

Hand Splints

- Can prevent deformity
- Added weight – benefit vs. Cost analysis
- Answer is individual and therefore variable
- Neoprene - McKie, Benik, Oval 8
- Thermoplast – can be various thickness and therefore weight (1/16", 1/8", perforated, tape)
- Kinesiotape may work -light

Don't Forget the Mouth!!

- **The Jaw:** important for hygiene, dental work, articulation and emergencies
 - Open wide
 - Side to side jaw deviation
- **The Tongue**
- **The Lips**



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See handouts for ideas you can customize

Technology Can Help

Alphabet taught to kids nowadays



A: APPLE



B: BLUETOOTH



C: CHAT:



D: DOWNLOAD



E: E MAIL



F: FACEBOOK



G: GOOGLE



H: HEWLETT
PACKARD



I: Iphone



J: JAVA



K: KINGSTON



L: LAPTOP



M: MESSENGER



N: NERO



O: ORKUT



P: PICASSA



Q: QUICK HEAL



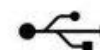
R: RAM



S: SERVER



T: TWITTER



U: USB



V: VISTA



W: WIFI



X: Xp



Y: YOU TUBE



Z: ZORPIA

It All Starts With Communication

Low Tech Options

Partner Assisted Auditory Scanning

- Can start ~ 9 months
- Set up a signal, “look at me if you want more bubbles”
- Progress to teaching via choices, “I have bubbles, TV and puzzles. Do you want bubbles? Do you want TV?”

Aided Language Stimulation

-teaching kids that symbols have meaning

More Communication

Higher Tech Options

- **Ipads/Iphones**

- Can use with a switch interface box if not able to use hands to swipe and click. Functional built into settings.
- Some apps have built in scanning or you can use IOS scanning with any app.

Handout: www.pinterest.com/bobbijohnsonaac

- **Home Automation**

- Amazon Dot and Echo now will recognize voice of AAC systems (i.e. Tobii)

Resources for Hardware

- Switch Interfaces
- Key guards and keyboard accessibility
- Mounts
- Switches
- Cases
- Stylus
- Environmental Control
- Websites
- Mobile Arm Supports

WWW.PINTEREST.COMBOBBIJOHNSONAAAC

Classroom Communication

Double Robot

Vgo





www.symbaloo.com/mix/bobbijohnsonaac

Wilmington Robotic Exoskeleton (WREX)

- Most appreciated by persons with mid range strength
- Provides support to arms and legs to allow movements while removing the effects of gravity.
- By adjusting the point of “balance” the arm is more mobile.
- External power of elastic elements
- Aids in: Feeding, Raising hands, Typing, Playing, Drumming, Fine motor skills

Playskin Lift and Playskin Air TM

- Garment based orthosis
- Provides varying levels of assistance to young infants with arm weakness who cannot yet reach or move their arms well
- Less bulky than other upper extremity systems
- **Tip of the ice berg....new versions coming!**

Different Models of Delivery

- Birth to Three
- Medical Model
- School Based Services
- Adjunctive/alternative

Birth To Three (early intervention)

- Federal Legislation: **IDEA - Part C**
 - Provides Funding and Criteria for states, which delegates to individual counties.
- EI services must be provided in **Natural Environment** to the *extent appropriate*
- Can bill insurances
- Can have parental cost sharing

Birth To Three Model Definitions

- Multi-disciplinary
- Interdisciplinary
- Transdisciplinary
- Primary Service Provider

0-3 Coaching Model

A style of interaction vs. a model of service delivery

School Based Therapy Services Educational Model

Ages 3 -21

- required by federal and state laws

Provided at no cost to the parents for students eligible for special education services

OT and PT are related services

- educational team must determine there is a need to support school performance

School Based Therapies

- Full team meetings (M-teams) every 3 years minimum.
- IEP developed with goals and reviewed annually.
- Be specific and include therapies when home bound, specific assistive tech, seating, transportation, etc.

Medical Model Service Delivery

- Provided in clinics, hospitals, homes, and community settings.
- The **physician, family and therapist** make decision re: the amount, frequency, and duration of therapy recommended.
- Services are paid for by medical insurance or private pay
 - insurance may dictate coverage and exclusions.
- Often obliged to not duplicate goals with other providers
 - 0-3 or school

Swimming and Aquatic Therapy

- Buoyancy allows movement with less effort (encourages a wider range of movement)
- Enhanced tactile input
- Reduces pain and tension in muscles
- Variety of supports available -head control
- Ping pong balls in panty hose to support limbs in water



Hippotherapy

- Horse used as treatment modality
- Specially trained PT or OT properly positions child on horse and directs horse, analyzes response
- Improve posture, balance, mobility and function
- Watch weight of helmet – may need neck support

Alternative Therapies

- **Massage**
- **Acupuncture**
- **Cranio-sacral**
- **Chiropractor**

**Choose a practitioner that is well informed
about Spinal Muscular Atrophy!!**

Changing Landscape



Disease Altering Therapies

- Spinraza
- Gene therapy
- Troponin skeletal muscle activators
- Other pharmacological therapies
 - Coming soon....

Impact on Therapy

- New goals and plan of care required
- New problems
- Change in decision making/timing of interventions
- Difficulty in predicting outcomes



What is Changing

- Large variability in response to treatment
 - EVERYONE agrees on this
- Long term changes unknown
- Need for ongoing assessment/reassessment
ESSENTIAL!

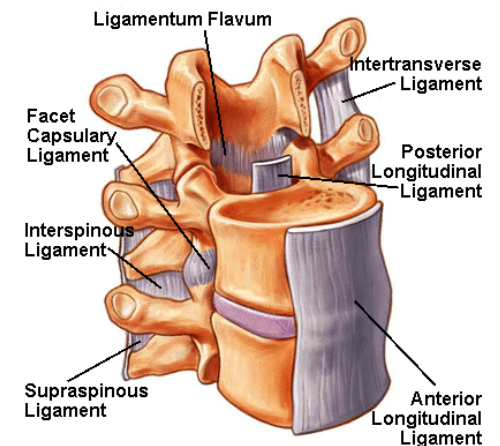
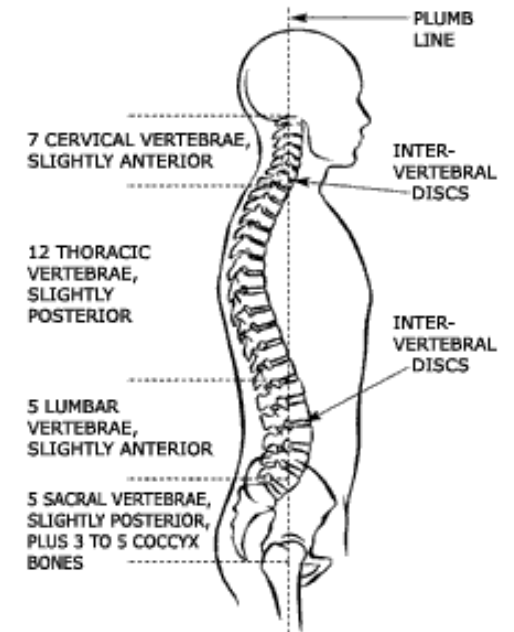
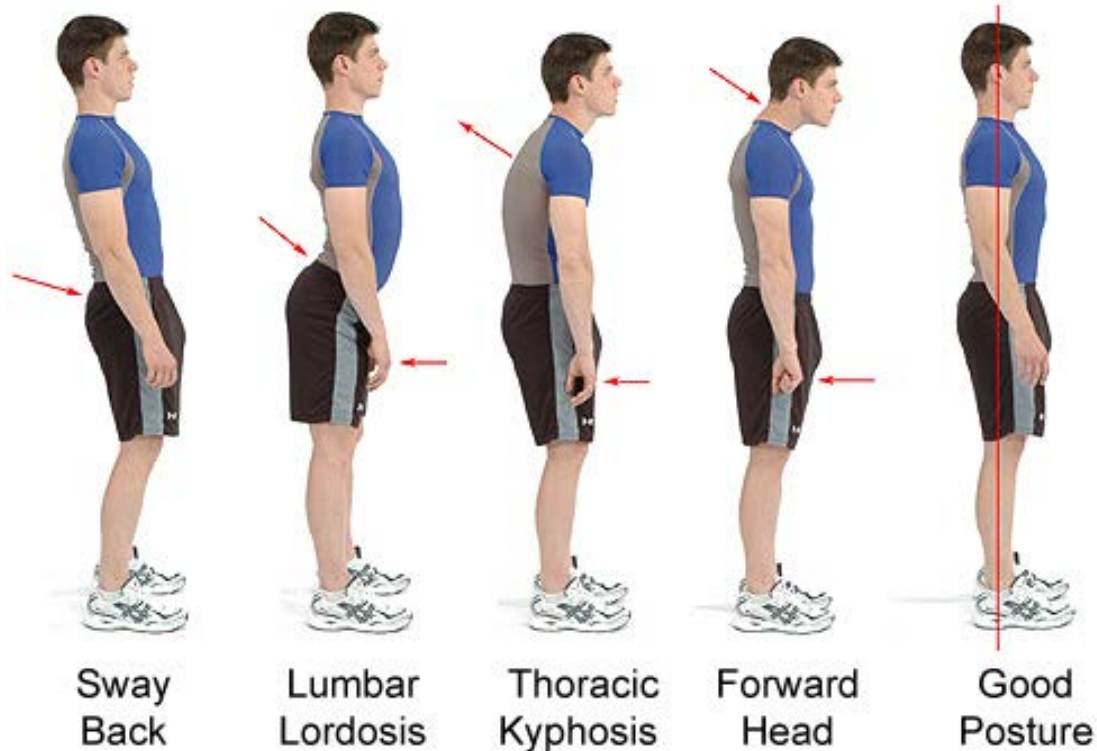
Tips, Tricks and Techniques for the Caregiver

According to the American Nurses Association (ANA)

- Up to 38% of all nurses experience back injuries. (2006)
- Musculoskeletal disorders:
 - more than any other work-related injury or illness, are responsible for lost work time, the need for protracted medical care, and permanent disability among health care workers.
- It is estimated that 12% of nurses leave the profession annually due to back injuries, and 52% complain of chronic back pain (ANA, 2009a).

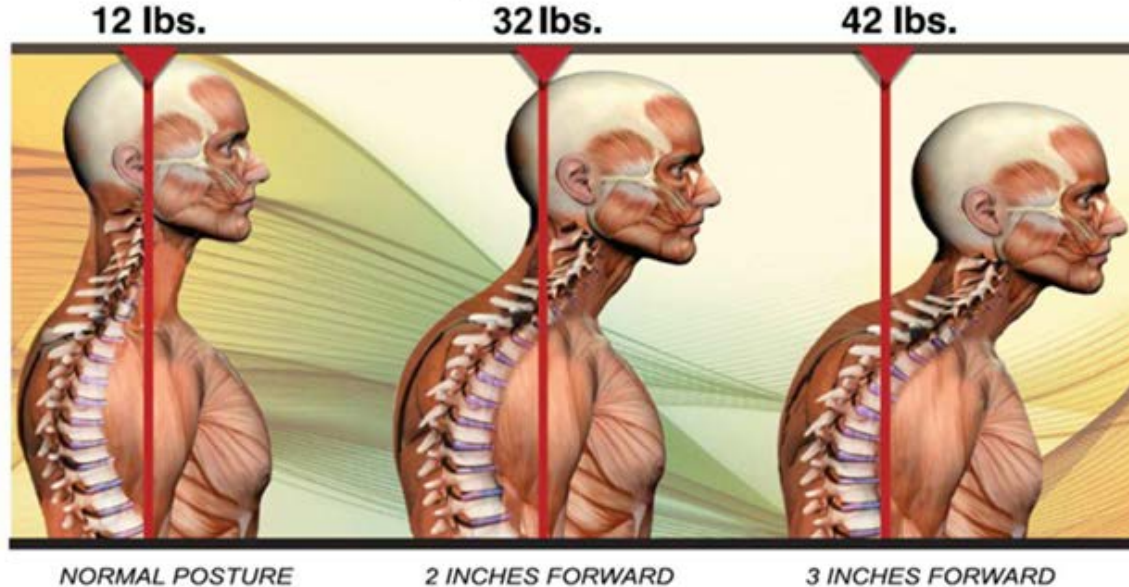


A Balanced Spine

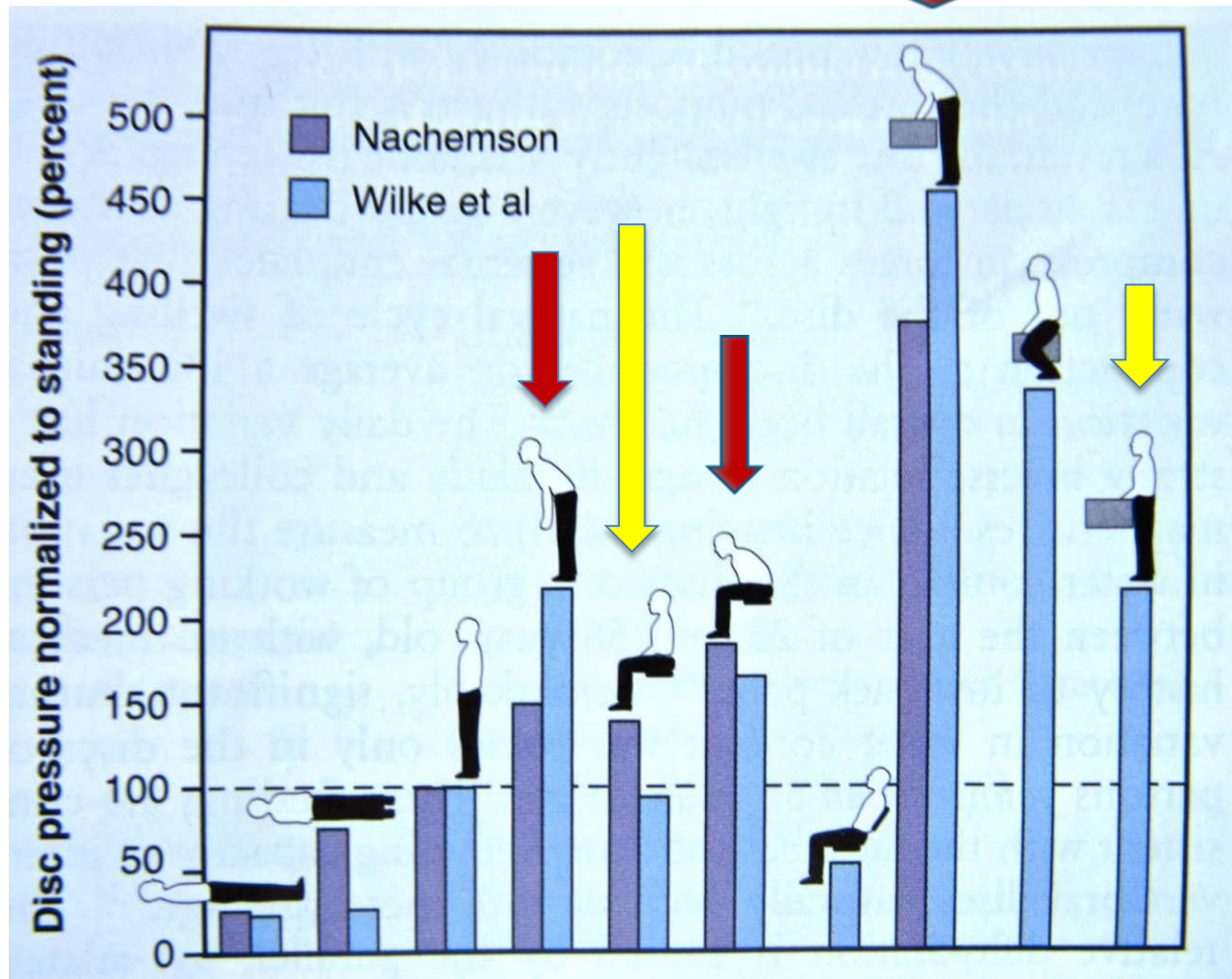


Posture Consequences:

How Heavy is Your Head?



Posture/lifting effect on Disc Pressure



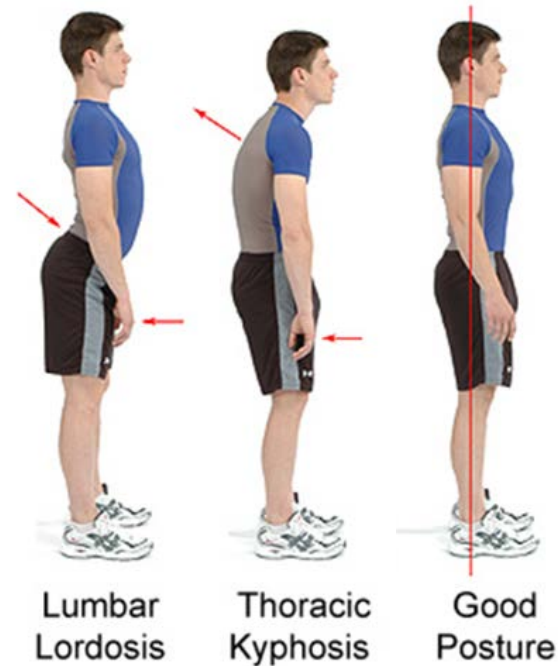
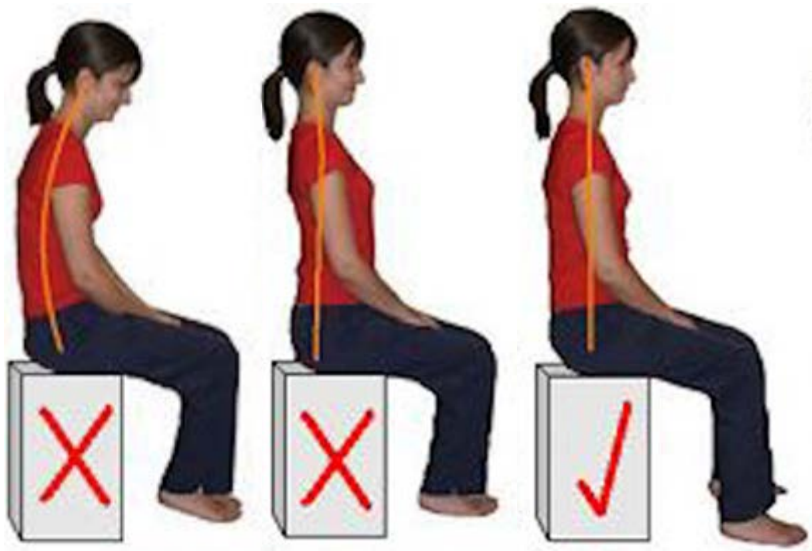
Using Good Body Mechanics

- Moving your body correctly is a skill that requires your constant attention.
- How well you perfect the skill can mean the difference between a fatigued or injured back and a healthy back.
- STOP ... THINK ... LIFT ... TRANSFER

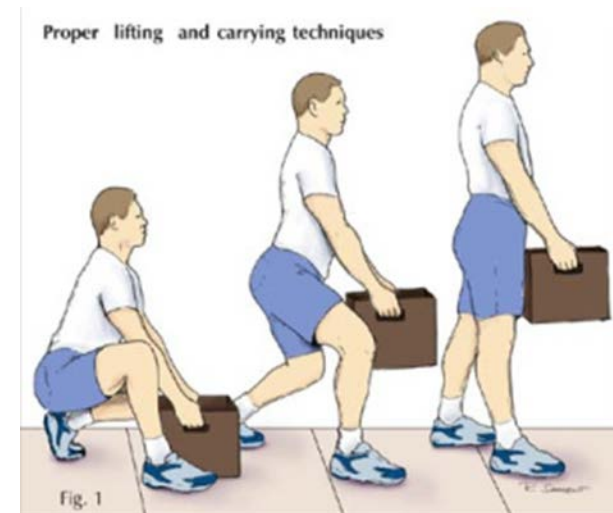
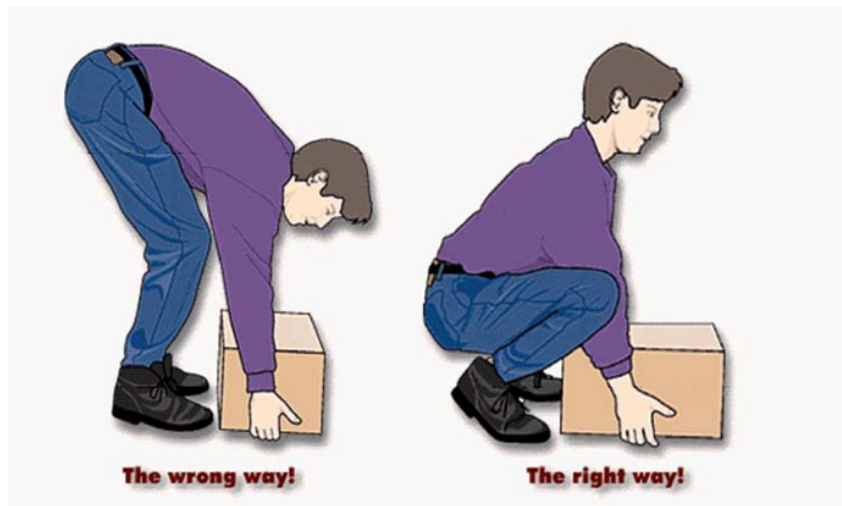
Tips and Techniques



Tighten abdominal muscles to help support your movements.



Bend **WITH** your knees/hips **NOT** your back



Keep your knees bent to make your legs work harder, reducing the stress on your back.

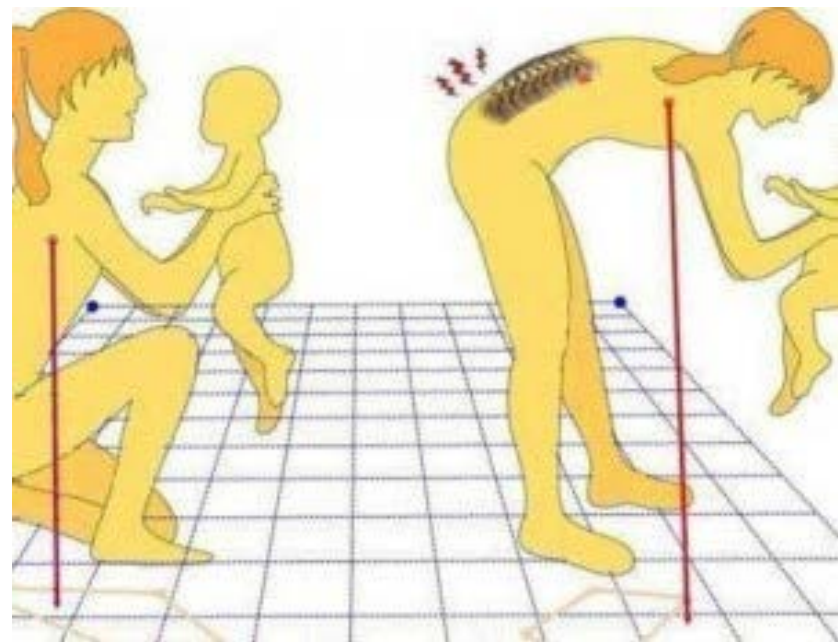
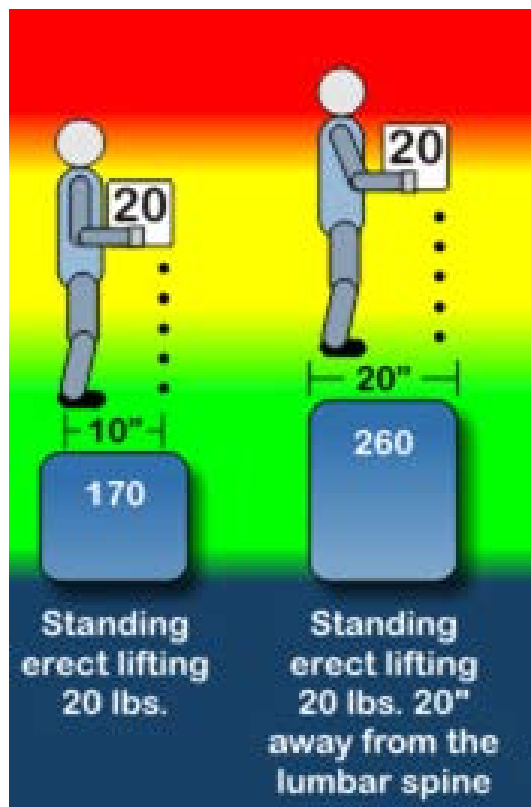


Neutral spine

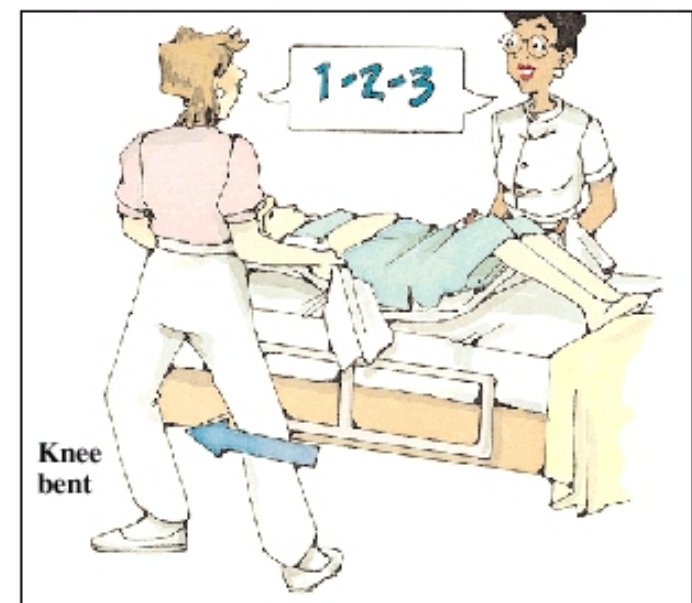
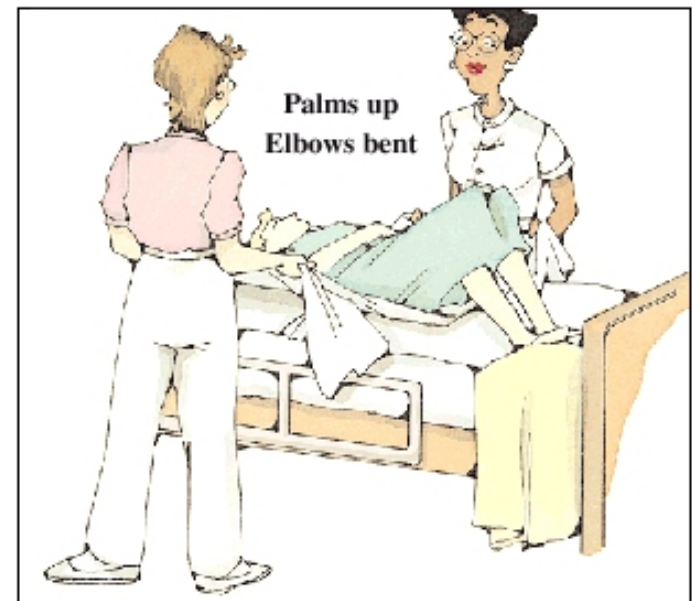
Flexed spine

Tip: Lift in stages to maximize legs.

Hold loads close to your body to minimize the effect of their weight.



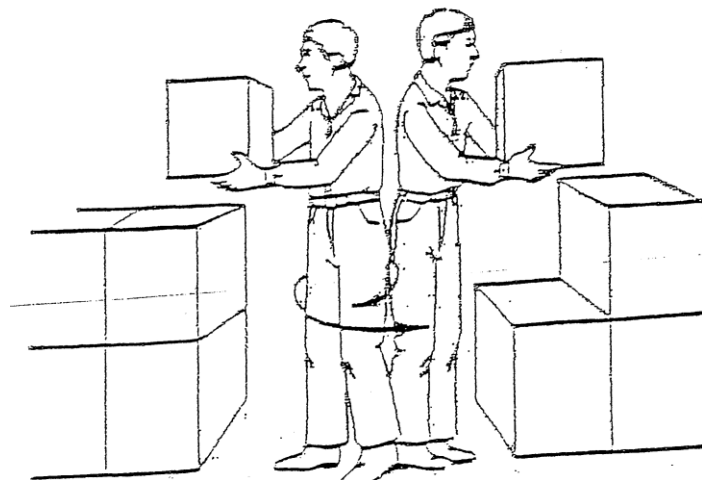
Tip: Draw
sheets!



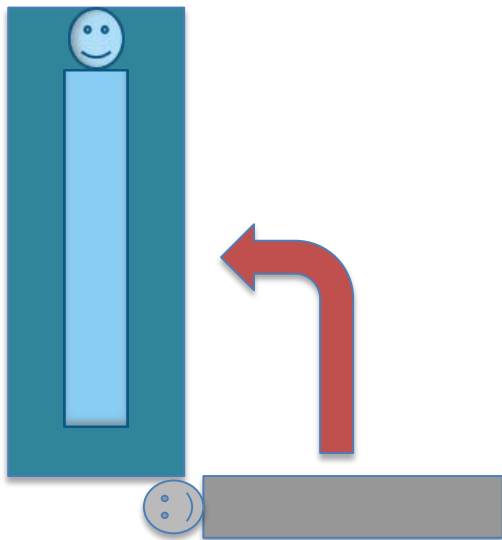
No BLT



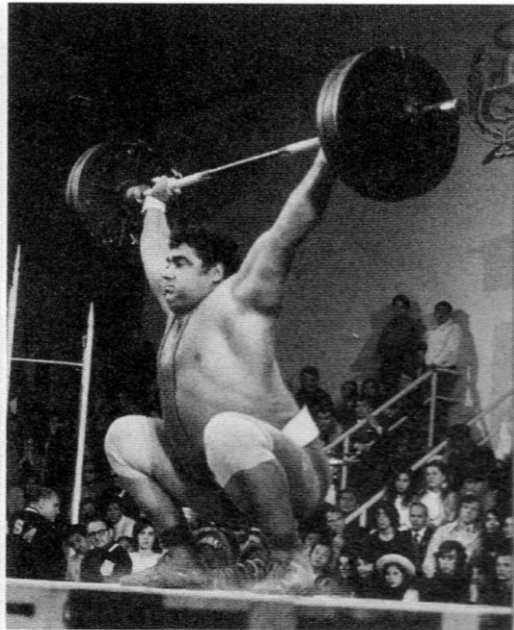
- **Bending, Lifting, Twisting** of your spine should never be done in combination.
- **Lift then move your feet:**
 - Make sure you turn your body as a whole, not just your back



TIP: Think
90 degrees
for transfers



Avoid quick, jerky movements.



Alexseev



Remember that each procedure for transfer and positioning has four main steps:

1. Plan the move and prepare the environment.
2. Starting position
3. Lifting Effort
4. Completing the move

Muscles

- Brace your abdominals to support your spine
- Remember your legs have the strongest muscles – use them

Environment

- Enough space and area free of obstacles
- If more than one person – work together and leader gives commands

You

- Stay fit and healthy
- Allow for rests
- Use proper technique
- Avoid/minimize lifting when tired or unwell
- Remember your muscles will get tired with repeated lifting

Set up

- Good posture – 3 curves
- Bend at your knees not waist
- Avoid twisting
- Hold load close to body
- Plan transfer

Equipment

- Use lifting equipment when possible
- Learn how to use correctly
- Store equipment in an accessible place

Questions, Discussion, Share



