

# Making Choices to Optimize Care and Quality of Life

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Make today a breakthrough.



**The Speakers have no relevant financial interests to disclose.**

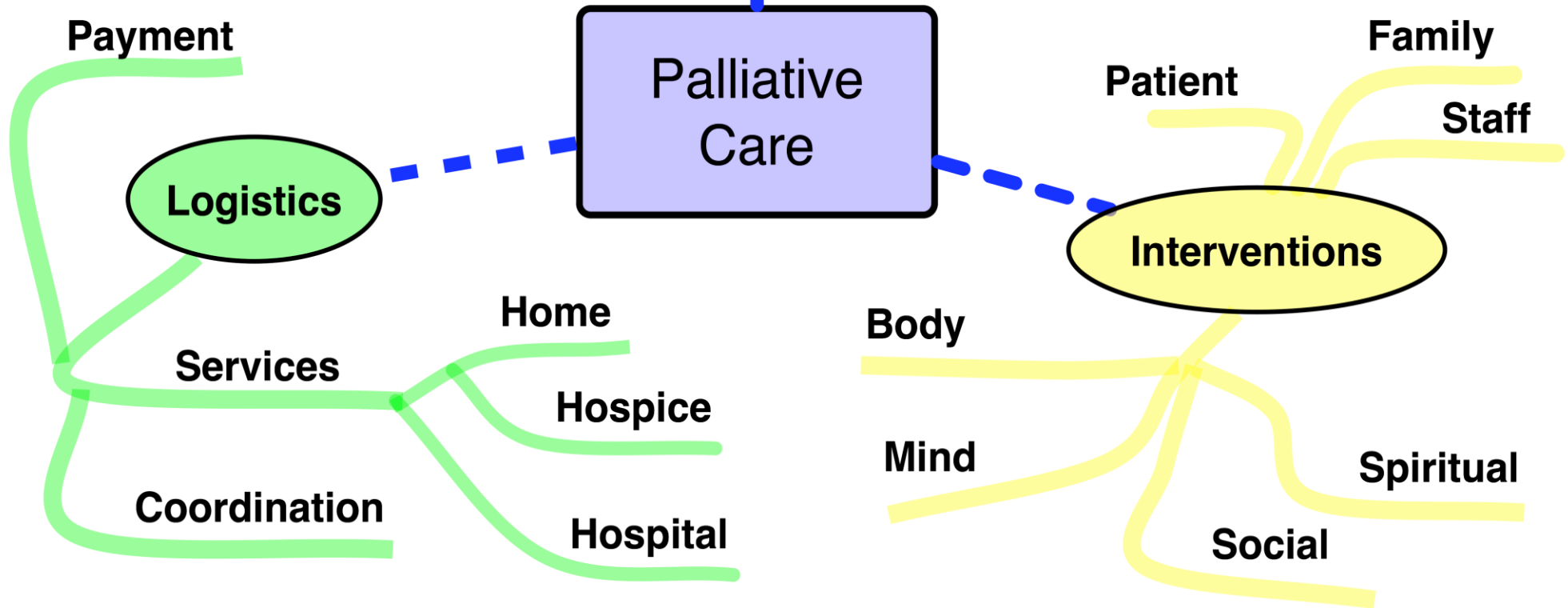
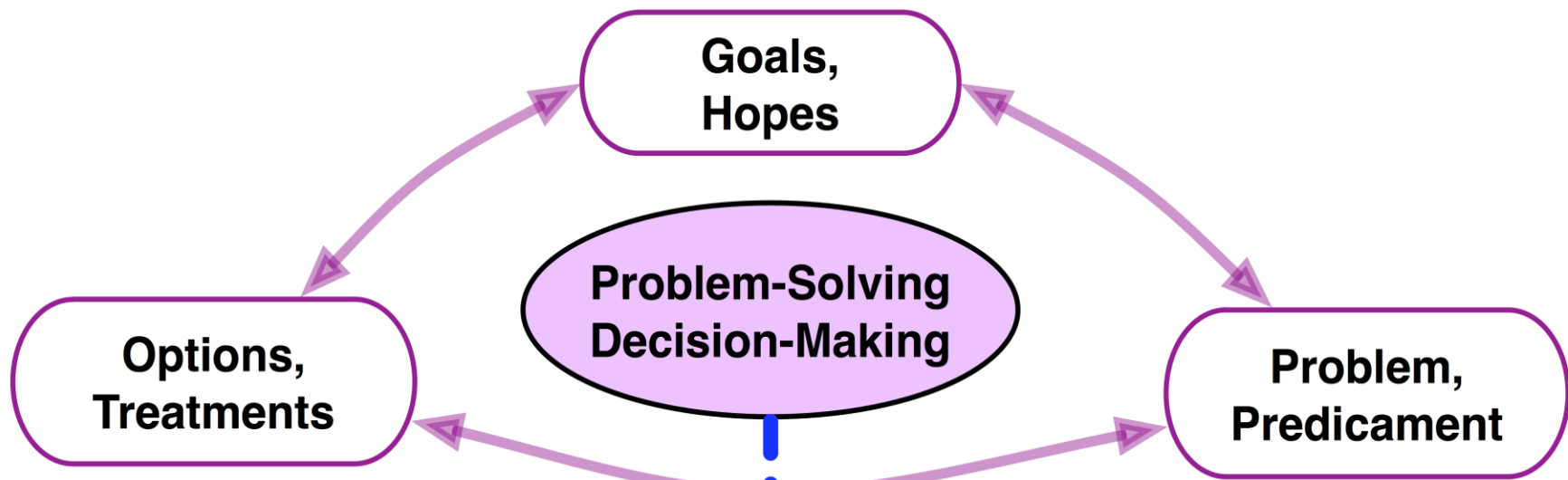
# Objectives for this Session

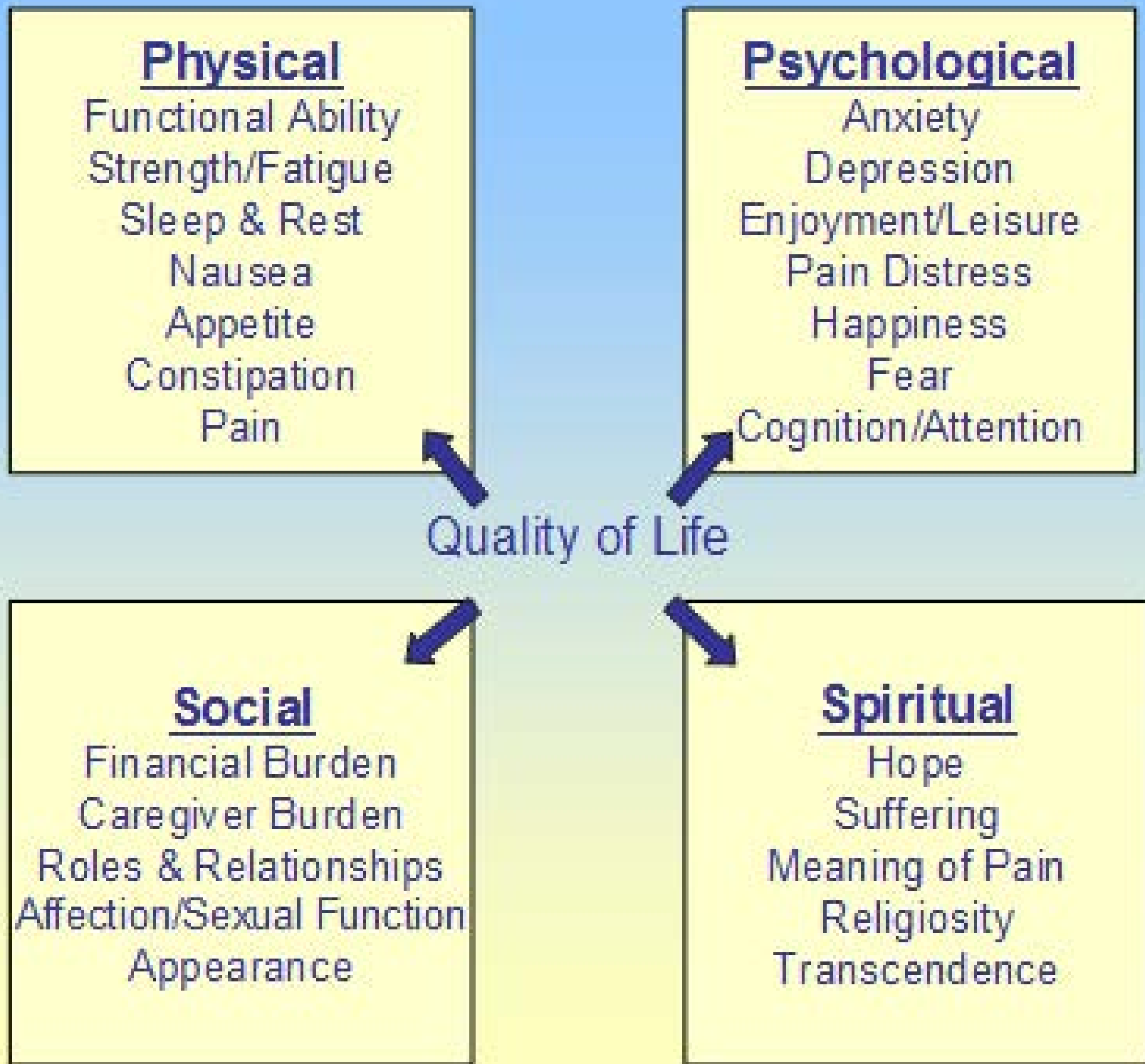
- Provide an overview of available options for care and ways to navigate choices
- Discuss making choices to optimize care & enhance quality of life
- Identify signs of pain, and explore various modalities to treat pain

# The Definition of Palliative Care

- Aims to improve quality of life of patients facing life-threatening illnesses, and their families, through the prevention and relief of suffering by early identification and treatment of pain and other problems, whether physical, psychological, social, or spiritual.

WHO, 2002





Adapted from Ferrell, et al. 1991

# What Do Families Need?

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- **Suffering:** Children even in the most technologically advanced medical centers in the world continue to suffer from pain and other symptoms
- **Decision-making:** Decisions that families are faced with in an era of increasing medical technology are more and more complex and require active support
- **Care coordination and transition:** Children's care is becoming increasingly complex, requiring care coordination
- **Family Support:** Families, including siblings, need continuous, long term compassionate, support

# Integrating Supportive Care

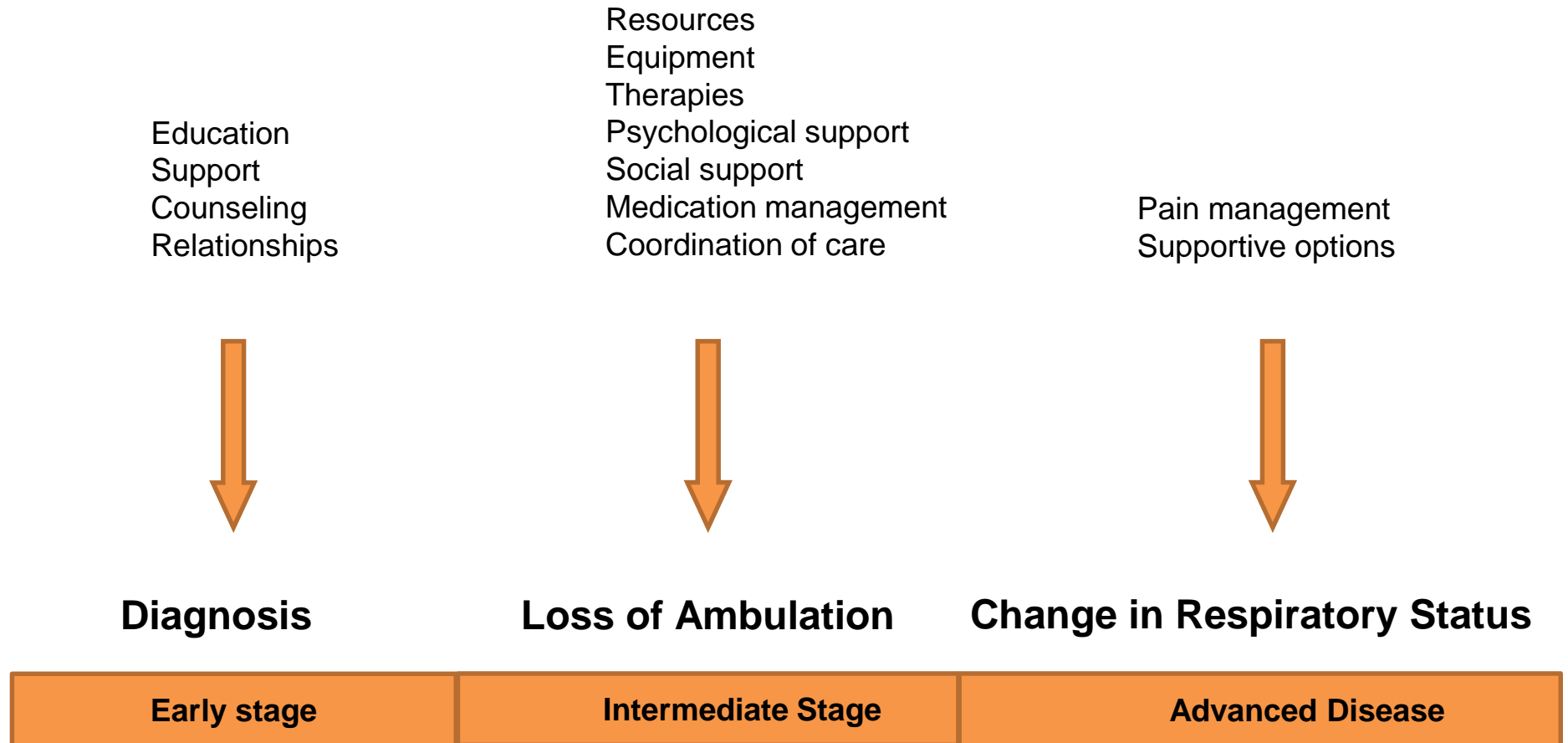
- 4 transition points at various disease stages
  - Peri-diagnostic
  - Treatment decision-making
  - Life-threatening events
  - End-of-life

Rushton, Erby, Cohn, & Geller, 2012



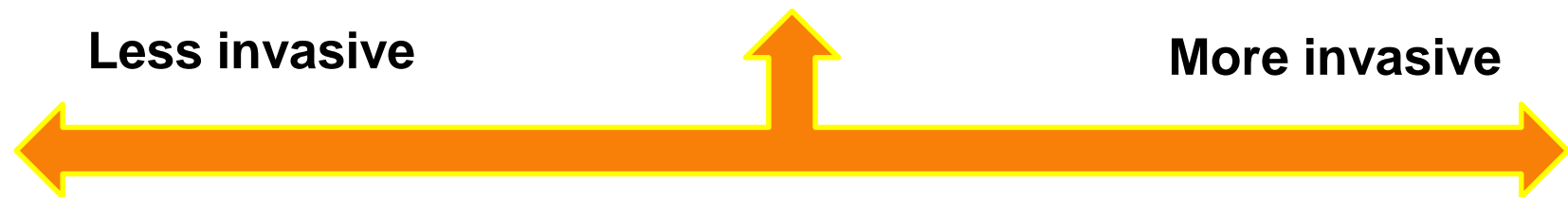


# A Model for Spinal Muscular Atrophy



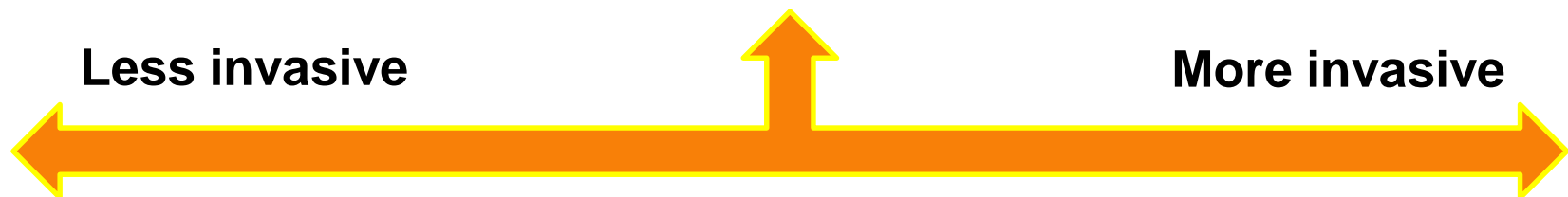
# Respiratory Care

- Non-invasive  
e.g.- CoughAssist™, BiPAP, Mechanical Ventilator
- Invasive  
e.g.- Tracheostomy + Mechanical Ventilator



# Nutritional Care

- Tube feedings:
  - Nasogastric (NG) tube
  - Gastrostomy (G) tube
  - G-tube and Nissen fundoplication
  - Gastrostomy-jejunostomy (G-J) tube



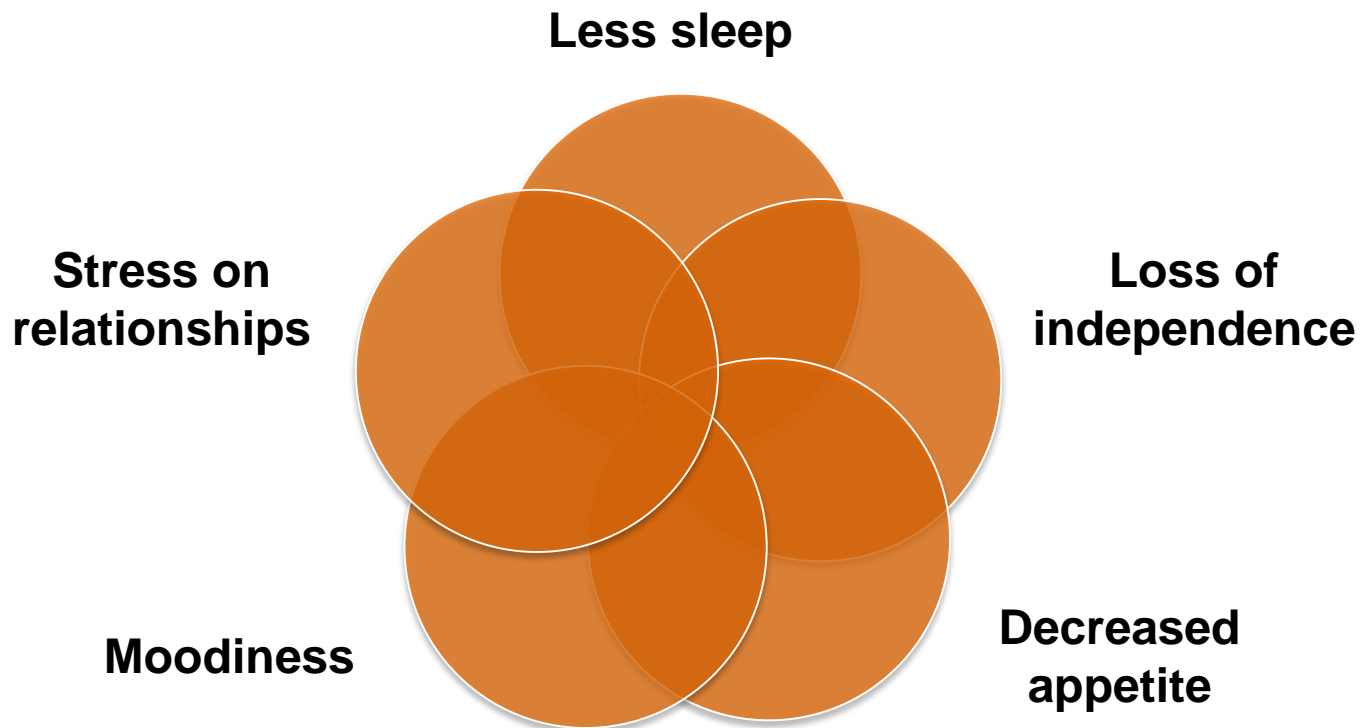
# Case Example

- **5 month old baby boy with SMA type I**
  - Early diagnosis
  - Family doesn't want "anything" done
  - Ask about BiPAP
  - Gets admitted and gets a G-tube
  - Parents don't want to talk about tracheostomy



WHAT DO WE DO?

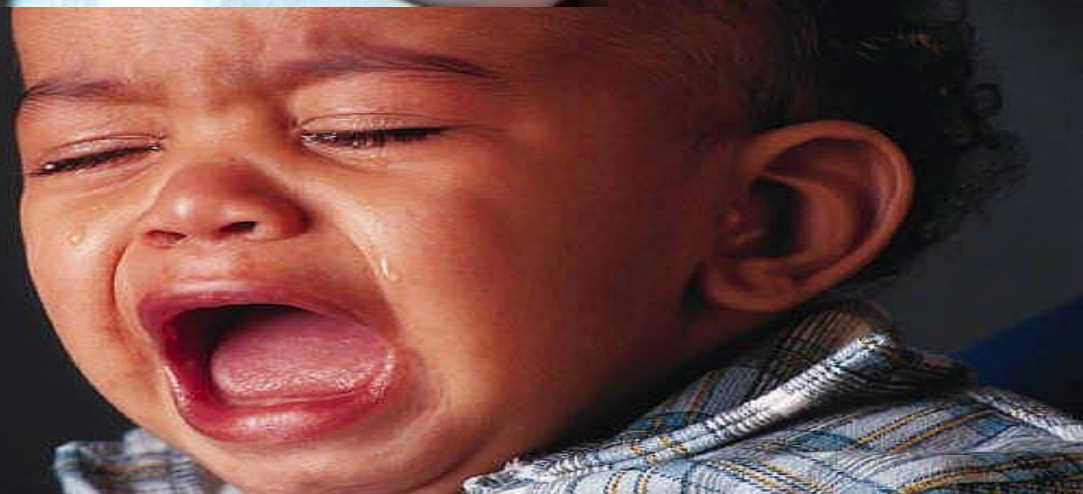
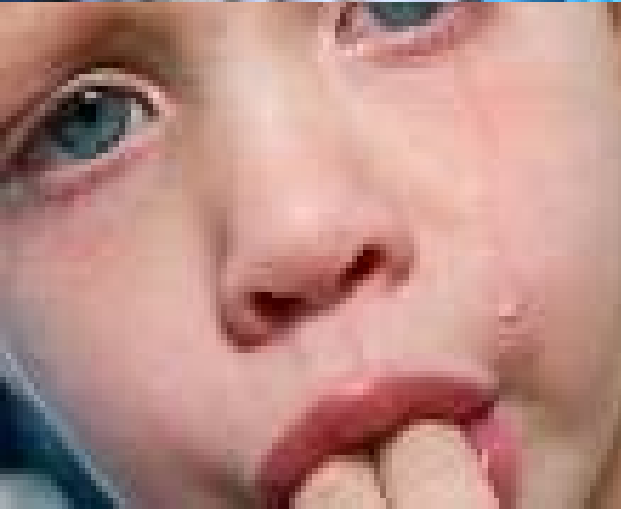
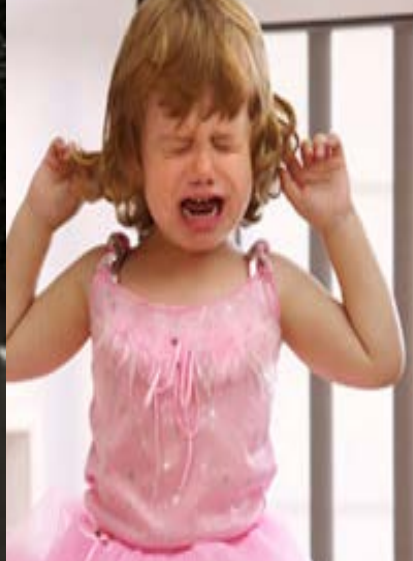
# Untreated Pain Directly Affects Quality of Life



# A Few Caveats on Pain

- Affects all aspects of life
- Multi-factorial (physical, psychological and emotional)
- Finding the source can be difficult and complicated
- Untreated pain can cause damage, affecting function and quality of life
- Learning skills to cope with chronic pain is essential
- Children may not have the words/language skills or ability to adequately describe pain, and thus their pain is often under treated







# General Concepts about Pain

# Myths Regarding Pain in Children (false beliefs)

- If you can distract a child, s/he does not have real pain
- Sleeping children are not in pain
- Behavior is directly related to pain intensity
- Pain builds character
- Infants have a higher threshold for pain
- Children will tell you when they are in pain
- Medications are always the best solution
- Medications are never the best solution

# Considerations for Non-Verbal Children

- ***Always*** consider other common sources of pain:
  - Constipation
  - Corneal abrasion
  - Dental pain
  - Ear infections
  - Fractures
  - Gastrointestinal reflux
  - Skin breakdown
  - Urinary tract infections (UTI's)

## Common Types of Pain

	<b>Acute (usu MSK)</b>	<b>Chronic (persistent)</b>	<b>Neuropathic</b>	<b>Procedural (episodic)</b>	<b>Referred</b>
	Normal pain reaction to injury that gets better	Injury heals but pain continues. Pain lasts beyond expected recovery time	Pain caused by damage to the nerves	Pain from procedures	Originates in one part of the body but is felt in another
Time-frame	Short lived, < 3 months	Generally, > 3 months	Can be acute or chronic	Short lived	
Changes we see	<p>↑HR</p> <p>↑BP</p> <p>↑Perspiration</p>	<p><b>Appetite changes</b></p> <p><b>Sadness, Fatigue</b></p> <p><b>Moodiness</b></p>	<p><b>Tingling,</b></p> <p><b>Burning</b></p> <p><b>“pins and needles”</b></p>	<b>Fear, Anxiety</b>	

# Possible Causes of Pain in SMA

- Musculoskeletal
  - Joint pain
  - Scoliosis
  - Hip subluxation  
(top of leg bone=ball of joint is partly out of socket)
  - Contractures – all joints but often knees/hips  
(shortening of muscles, tendons or ligaments)
  - Muscle cramping / muscle spasm
- Fractures
- Positional
- Equipment (braces, wheelchairs, respiratory equipment, etc.)
- Immobility
- Pressure sores
- Pain after procedures (G-tube, scoliosis surgery, etc. )

# Possible Causes of Pain in SMA

	Pain Considerations	Treatment	Medication
<b>Scoliosis</b>	<ul style="list-style-type: none"> <li>- Curvature can cause irritation of nerves</li> <li>-Can lead to leg pain and affect breathing</li> </ul>	<ul style="list-style-type: none"> <li>PT/OT</li> <li>Bracing</li> <li>Surgery</li> </ul>	<ul style="list-style-type: none"> <li>Non-Steroidal Anti-Inflammatory</li> <li>Gabapentin</li> </ul>
<b>Contractures</b>	<ul style="list-style-type: none"> <li>Muscle imbalance</li> <li>Muscle weakness</li> <li>+/- Spasticity</li> </ul>	<ul style="list-style-type: none"> <li>Orthotics</li> <li>PT/OT PROM</li> <li>Repositioning</li> <li>Moist heat</li> </ul>	<ul style="list-style-type: none"> <li>Non-Steroidal Anti-Inflammatory</li> <li>Gabapentin</li> </ul>
<b>Hip Dislocation</b>		<ul style="list-style-type: none"> <li>Close observation</li> <li>Possible surgery</li> </ul>	

# TIPS for Parents: Keep a Pain History

- Quality or nature of pain (what was happening when it started)
- Mode of onset and location (sudden, gradual, ongoing)
- Duration and chronicity (how long and how often)
- What makes it worse or better
- Description (throbbing, burning, dull, sharp, radiates )
- Timing of pain (does it happen at set times)

# Tips for Parents: Pain Medication

- Don't wait for pain to be severe before treating
- Give at regular intervals to prevent pain before the next dose
- No single medication or dose will work for all children
- Weigh risk of, and treat, potential side effects



# Non-Medication Pain Treatments

Age	Pain Behavior	Intervention
<b>Infant</b>	<ul style="list-style-type: none"> <li>•High pitch cry</li> <li>•Grimacing</li> <li>•Difficulty sucking</li> <li>•Avoiding eye contact</li> <li>•Inconsolability</li> </ul>	<ul style="list-style-type: none"> <li>•Swaddling</li> <li>•Position changes</li> <li>•Gentle massage</li> <li>•Pacifier dipped in sugar water</li> </ul>
<b>Toddler</b>	<ul style="list-style-type: none"> <li>•Sleep difficulties</li> <li>•Protect area of discomfort</li> <li>•Change in play/appetite</li> <li>•Irritable</li> </ul>	<ul style="list-style-type: none"> <li>•Distraction</li> <li>•Art/Music Therapy</li> <li>•Guided Imagery</li> <li>•Massage</li> <li>•Aqua therapy</li> </ul>
<b>School-age and Adolescent</b>	<ul style="list-style-type: none"> <li>•Guarding</li> <li>•Insomnia</li> <li>•Mismatched cues</li> <li>•Quiet or Subdued</li> <li>•Irritable or Anger</li> </ul>	<ul style="list-style-type: none"> <li>•Breathing techniques</li> <li>•Guided Imagery</li> <li>•Acupuncture (&gt;8 years)</li> <li>•Hypnosis</li> <li>•Art/Music Therapy</li> </ul>

# Hot / Cold Therapy

Cold Compress	Decreases blood flow, decreases inflammation and pain	Injury, bruise, swelling	Can make tension and spasms worse  10-15 minutes on/off
Warm Moist Heat	Increases blood flow, relaxes sore muscles, decreases spasms	Chronic muscle pain, stress	Can make inflammation worse  10- 15 minutes on/off

# Integrative Therapies

Use of conventional therapy, as well as complementary therapies, together in an individualized plan of care.

# Guided Imagery

The deliberate use of the imagination to help the mind and body heal, stay well, or perform well.

# Therapeutic Touch and Massage

*“Therapeutic Touch is an intentionally directed process of energy exchange during which the practitioner uses the hands as a focus to facilitate the healing process.”*

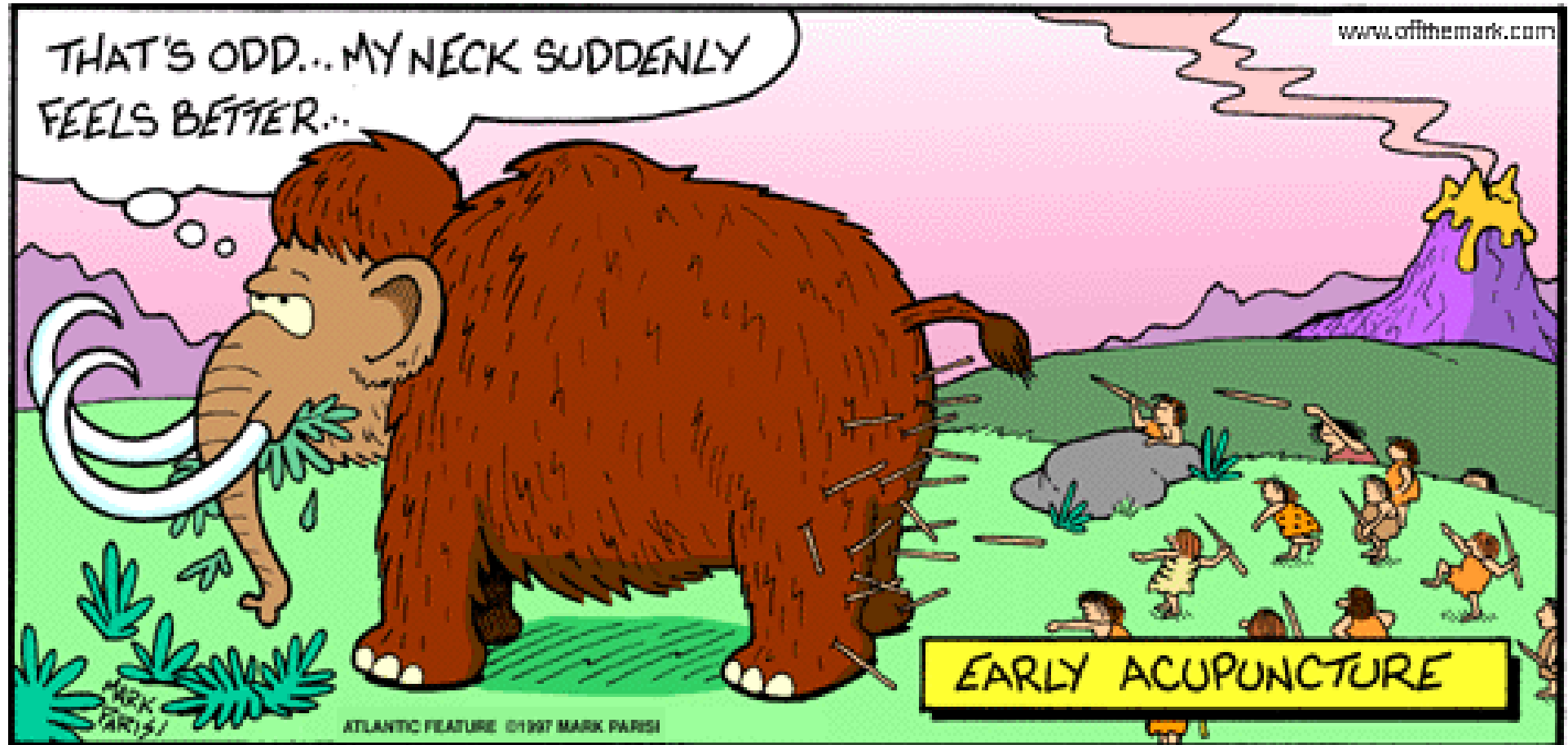
*Nurse Healers-Professional Association*

# Acupuncture

**off the mark**

by Mark Parisi

[www.offthemark.com](http://www.offthemark.com)



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# Case for Discussion: Abby

- Abby is a 9 month old infant with SMA type 1 who is home requiring BiPAP support 12-16 hours per day.
- She receives nutrition via a G-Tube.
- Her parents are concerned because she seems irritable off and on during the day for the past several days.
- They are worried she is having pain and wonder what to do next.

## Abby (Continued)

- Abby is now 2 years old and is at home with BiPAP support 20-24 hours per day.
- She has contractures and seems to have discomfort even when sitting in her customized chair.
- Her therapists and parents have tried many different positions, massage, and physical therapy.
- Her parents are concerned because she is having difficulty sleeping and seems uncomfortable for a few weeks.



# Older Abby

- Abby is now 5 years old and is at home with a tracheostomy and a ventilator.
- Her contractures have worsened and her family feels she is having positional pain.
- Her therapists and parents have tried many different positions, continued physical therapy, aqua-therapy, massage and Tylenol.
- Her parents are concerned because she is having ongoing difficulty sleeping and seems uncomfortable with care.

# SMA Conference Survey

Please complete your conference survey at this link:

<https://www.surveymonkey.com/r/2017SMAAnnualConference>

**Or fill out the paper survey in your conference folder.**

- **All participants who complete a survey by 10:30 am on Sunday July 2nd, will have their name entered into a raffle for a brand new iPad!**
- **The winners will be drawn and announced on Sunday, July 2nd at the Closing General Session/It's a Wonderful Life.**