

# Type I- Optimal Care for SMA

2016 Annual SMA Conference  
Disneyland Hotel, Anaheim, CA  
June 16, 2016



**Make today a breakthrough.**

# Panel

- Mary Schroth, MD
- Garey Noritz, MD
- Rebecca Hurst Davis, MS, RD, CSP, CD
- Vanessa Battista, RN, MS, CPNP
- Karen Patterson, MS, PT, PCS

# Breathing



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- **Mary Schroth, MD**

- University of Wisconsin School of Medicine and Public Health

- American Family Children's Hospital  
Madison, Wisconsin

- mschroth@wisc.edu

- Paid Medical Professional Education Consultant

- mary@curesma.org

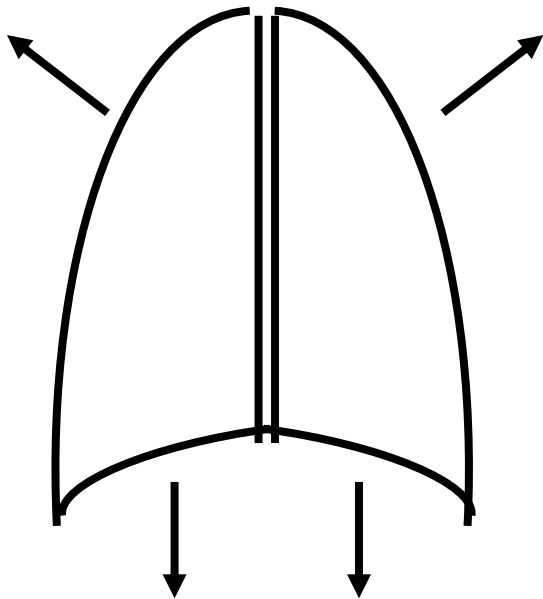


# SMA Breathing Basics

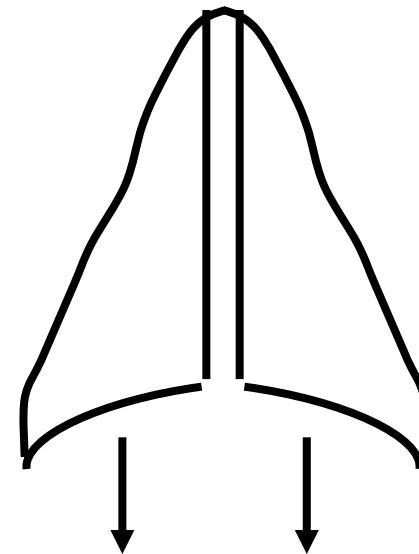
- Very weak muscles between the ribs
- Chest is very soft and flexible
- Diaphragm is primary muscle used to breath

# Chest Wall Changes

**Normal**



**SMA**



# Results of Respiratory Muscle Weakness in SMA

1. Difficulty coughing
2. Small shallow breaths during sleep
3. Chest wall and lung underdevelopment
4. Frequent pneumonias that contribute to muscle weakness.

# Loosen Secretions

- **Manual Chest Physiotherapy or Mechanical Percussion**



- **Postural Drainage**



*Do before eating or at least 30 minutes after eating*



# Machine for Coughing



Respironics  
Cough Assist™ CA-3000



Hill-Rom Vital Cough™



Respironics  
Cough Assist™ T70

# Machines to Support Breathing

- Bi-level positive airway pressure (BiPAP)
- Home ventilator



# Recommendations

- If difficulty breathing, lay flat.
- Car Bed rather than Carseat for travel.
- Request consultation with pulmonologist locally.
- Immunize your child.
- Request Synagis during the winter months to prevent RSV infection.

# At the Conference

- Specific Sessions regarding Respiratory Care:
  - Saturday:
    - 8:45 am-10:15 am: [Breathing Basics and Care Choices for Type I](#) – South Ballroom B
    - 10:30 am-12:00 pm: [Family Readiness for Emergencies](#) – South Ballroom B

# Supportive Care



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# Supportive Care

- Garey Noritz, MD
  - The Ohio State University,
  - Nationwide Children's Hospital, Columbus Ohio
  - [Garey.Noritz@nationwidechildrens.org](mailto:Garey.Noritz@nationwidechildrens.org)

# At the Conference

- Specific Sessions regarding supportive care:
  - Friday
    - 9:00-10:15 am:
      - Siblings Talk It Out (12-17 years old) – Adventure
    - 10:30 am-12:30 pm:
      - Sharing your Type I experience – North Ballroom A
      - Grandparents Talk It Out Type I – Monorail A & B
    - 2:00-3:30 pm:
      - Yoga Therapy for SMA – South Ballroom B
  - Saturday
    - 10:30-12:00 pm:
      - Recognizing and Managing Pain in SMA – North Ballroom
    - 1:30-3:00 pm:
      - Dad's Time: A Workshop for Father's Only - Type I – North Ballroom B
      - Siblings Talk It Out (Ages 5-11) – Monorail A & B

# Nutrition for Children with Spinal Muscular Atrophy Type I



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# Nutrition

- Rebecca Hurst Davis, MS, RD, CSP, CD
  - Intermountain Healthcare
  - [becky.hurst-davis@imail.org](mailto:becky.hurst-davis@imail.org)

# Objectives

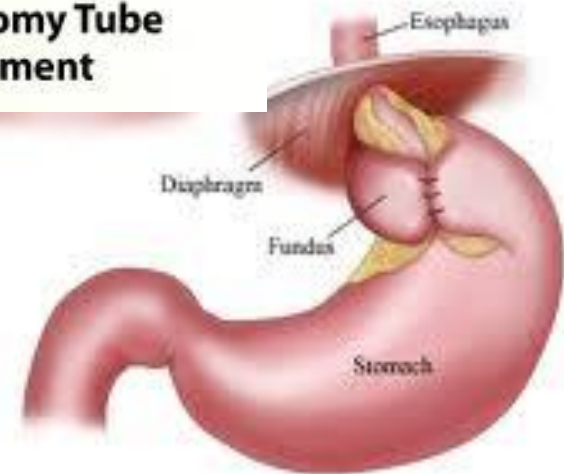
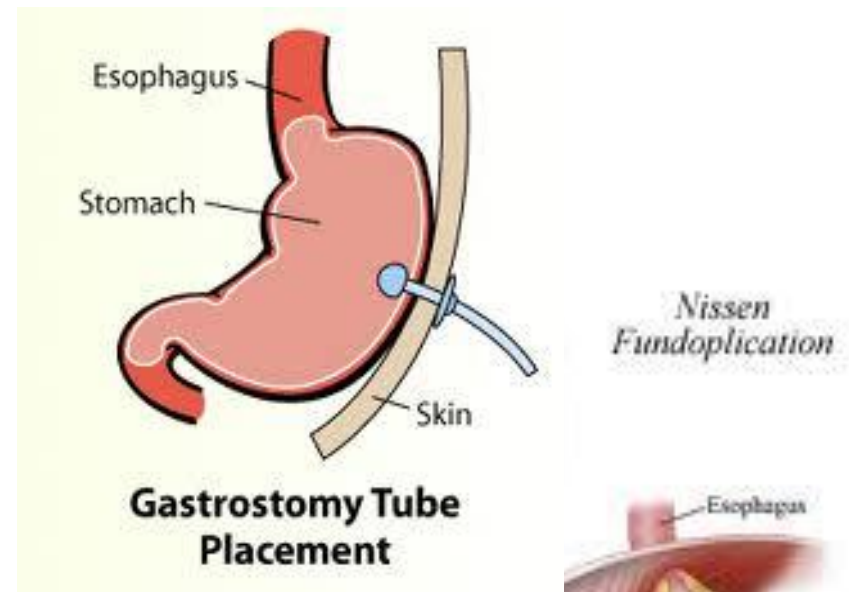
- Highlight the most important nutritional issues for children with Type I SMA
  - Safe Feeding/Feeding Tubes
  - Nutrition Referrals
  - Illness
  - Diet

# Safe Feeding/Feeding Tubes

- Intake enough nutrients to grow, and maintain growth without aspiration.
- For proactive care: Children with Type I will eventually need a feeding tube
- Recommend a swallow study at diagnosis and routinely after
- Nasojejunal (NJ) feedings short term (tube in the nose can affect breathing)
  - Acute illness
  - Interim while waiting for g-tube surgery

# Feeding Tubes

- Gastrostomy (G-tube) common for long term nutrition support
- Nissen fundoplication
- Speak with GI doc



# When to place a G-Tube

- If no longer safe to swallow.
- Child's growth is slowing, starting to have feeding troubles.
- Proactive Approach: Start to plan for tube shortly after diagnosis of SMA Type I.
- Work with a knowledgeable RD from the beginning
  - EVEN if you are not having nutrition problems!
    - Aids in maintaining a satisfactory nutrition state
    - Builds a relationship with local health providers
    - Gives guidance to improve nutrition
    - Helps monitor for safe feeding

# Nutrition Referrals

- Speech Therapy evaluations and intervention
  - Assess for swallowing issues,
  - Improve oral motor skills,
  - Alter texture of foods to decrease fatigue, thickeners, pureed foods,
  - Nipple flow/ bottle changes for bottle-fed infants
  - Positioning

# Nutrition Referrals

- Dietitian evaluation (every 3-6 months)
  - Evaluate growth: length, weight, head growth, other body measures over time
  - Reviews:
    - Feeding schedule
    - Overall diet: Analyzes 3 day diet record or reviews 24 hour recall
    - Fluid
    - Vitamin and mineral needs
    - Diet tolerance
  - Makes recommendations or changes in “milk”, formula preparation, feeding schedule, and supplements to meet nutrient needs.
  - Works with MD to improve your child’s health and QOL.

# Illness-No Prolonged Fasting

- Children with Type I should not fast longer than 6 hours- especially when ill!
- Reduced muscle mass
  - Less nutrition reserves for protein, carbohydrate and mineral metabolism.
- Prolonged fasting, diarrhea, vomiting, and fevers can be life threatening to a child with Type I SMA
- If your child does have diarrhea and/or vomiting, seek medical advice and attention.



# DIET

- Very individualized
  - child's tolerance
  - parental choice.
- Formulas, including the elemental “Amino Acid” diet have not been studied in SMA.
- Many formula types:
  - regular>soy>semi-elemental>elemental
- Clinical symptoms may indicate need for a more broken down formula and/or change in feeding schedule.
- Consider benefits of breast milk.
- Especially use dietitian if choosing AA diet.

# Supplementation

- May require a multivitamin or individual supplement such as iron, vitamin D. Check with MD or RD.
- Avoid Mega-doses of supplements
- Recommend routine nutrition labs/exams



Many shades of grey, there is not one umbrella SMA diet. Follow diet that works for your child and family. Diet may change as your child's health changes.

# At the Conference?

- Specific Sessions regarding Nutrition:
  - Saturday:
    - 8:45-10:15 am:
      - Getting the Most Out of What You Eat! Nutrition for Oral Feeders – North Ballroom A
    - 1:30-3:00 pm:
      - Tube Feeding and SMA: Recommendations and Practices – South Ballroom B

# Options for Care



**Make today a breakthrough.**

# Options for Care...



- Vanessa Battista, RN, MS, CPNP
  - The Children’s Hospital of Philadelphia
    - Pediatric Advanced Care Team (PACT)
  - [battistav@email.chop.edu](mailto:battistav@email.chop.edu)

# World Health Organization Definition of Pediatric Palliative Care

- Aims to improve quality of life of children 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

# What Does Palliative Care Mean for My Family?

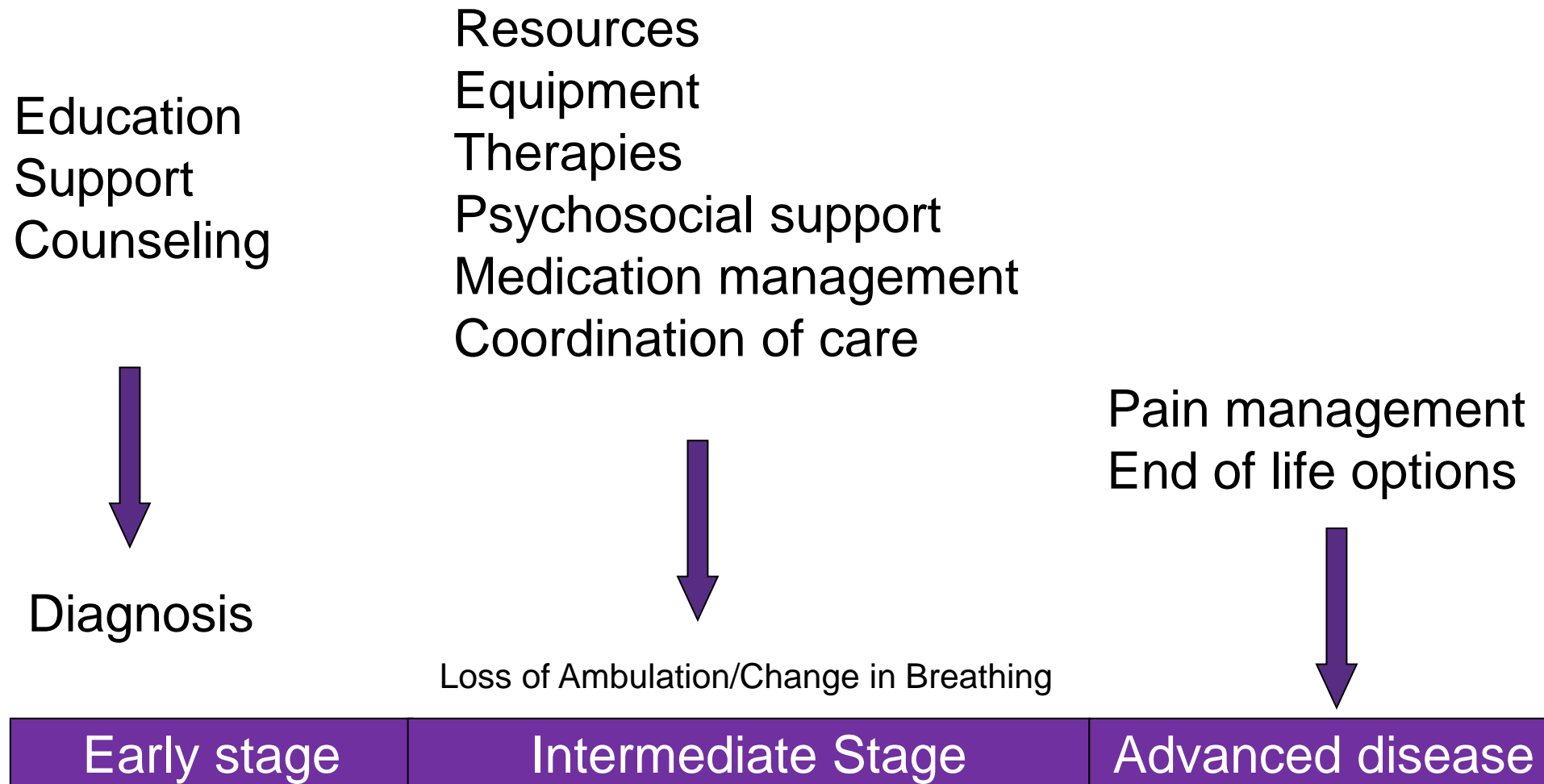
- Begins at diagnosis
- Upholds quality of life
- Involves the whole family and community
- Continues with or without disease directed treatment
- Supports families in making the choices that are best for YOU
- Does NOT mean 'giving up', 'no care' or 'no options'!!



# Maintain Care and Support

- Comprehensive interdisciplinary care
- Practical advice and support: inpatient and outpatient
- Define and respect quality of life for each child/family
  - physical, psychological, social, and spiritual preferences
- Help families think through decisions and make choices
- **Support for the whole family throughout a child's journey**

# Family-Centered Palliative Care for SMA



# At the Conference

- Specific Sessions regarding supportive care:
  - Friday
    - 9:00-10:15 am:
      - Siblings Talk It Out (12-17 years old) – Adventure
    - 10:30 am-12:30 pm:
      - Sharing your Type I experience – North Ballroom A
      - Grandparents Talk It Out Type I – Monorail A & B
    - 2:00-3:30 pm:
      - Yoga Therapy for SMA – South Ballroom B
  - Saturday
    - 10:30-12:00 pm:
      - Recognizing and Managing Pain in SMA – North Ballroom B
    - 1:30-3:00 pm:
      - Dad's Time: A Workshop for Father's Only - Type I – North Ballroom B
      - Siblings Talk It Out (Ages 5-11) – Monorail A & B
      - Ethics of Clinical Research – Center Ballroom

# Therapy



**Make today a breakthrough.**



- Karen Patterson, MS, PT, PCS
  - University of Wisconsin-Madison Doctor of Physical Therapy Program
  - Faculty Associate
  - CureSMA Medical Advisory Board
  - [pattersonk@pt.wisc.edu](mailto:pattersonk@pt.wisc.edu)

# At the Conference?

- Talk to other Families
- Establishing your “Team”
- Talk to Equipment Vendors
- Write stuff down!



# Musculoskeletal and Rehab Team Members

- **Orthopedic doctor**
  - Bones and muscles
- **Physical Medicine and Rehabilitation doctor**
  - PM&R doc- oversees rehabilitation care
- **Physical Therapist (PT)**
  - strength, walking, sitting, posture, balance
- **Occupational Therapist (OT)**
  - adaptive aids, fine motor/hand skills
- **Speech Therapy (ST)**
  - language, jaw issues, feeding
- **Orthotist**
  - makes orthotics or braces

# What does “Therapy” focus on?

- We’ ll help you with:
  - Positioning during daily routines and activities to encourage participation
  - Adapting toys for play and exercise
  - Expanding mobility options
  - Using equipment effectively
  - Monitoring posture and alignment and the need for braces, surgery, seating, etc.



# Balance between play and therapeutic activities

**Interventions should:**

- **S**UPPORT function
- **M**AINTAIN abilities
- **A**UGMENT play and mobility



# Is Your Child Entitled to Services?

- **Different Models of Delivery of Services:**
  - Early Intervention (Birth to Three yrs of age)
    - Typically in the home
    - May not include all disciplines
  - School Based Services
    - Provided under IDEA (Individuals with Disabilities Education Act)
    - Therapy is a “related service”
    - Educational goals (access, participation)
    - Includes transportation and possible therapy at home if home bound
  - Medical Model
    - Typically outpatient
    - Check # of visits with insurance
    - May also receive inpatient services if hospitalized (for any reason)
- **Typically can't duplicate services**

# Various things you might see and wonder about over the weekend



- Specific Sessions regarding Therapy, Rehab and Orthopedics:
  - Friday:
    - 2:00–3:30 pm:
      - Aquatic Physical Therapy for Fun and Function - Type I – Neverland Pool
      - Standing, Walking and Mobility: Decision making and Options – Center Ballroom
      - Yoga Therapy – South Ballroom B
  - Saturday:
    - 8:45–10:15 am:
      - Orthopedic Management – North Ballroom B
    - 10:30 am–12:00 pm:
      - Aquatic Physical Therapy for Fun and Function – Type II, Type III and Adult SMA – Pool
    - 1:30 – 3:00 pm:
      - The Role of PT and OT in your Child’s Life – Being Part of Therapy Team – South Ballroom A

# SMA Conference Survey

**Please complete your conference survey  
at this link:**

<https://www.surveymonkey.com/s/2016AnnualSMAConference>

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

- **All participants who complete a survey by 10:30 am on Sunday June 19th, will have their name entered into a raffle for a brand new iPad!**
- **The winners will be drawn and announced on Sunday, June 19th at the Closing General Session/It's a Wonderful Life.**

