

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



Make today a breakthrough.

- **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

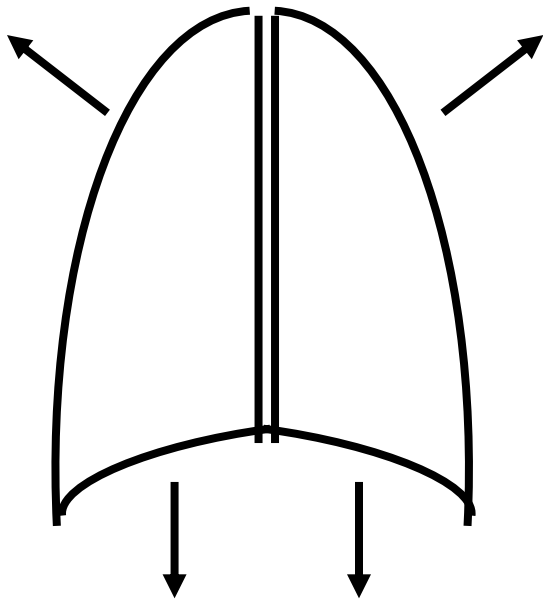


SMA Breathing Basics

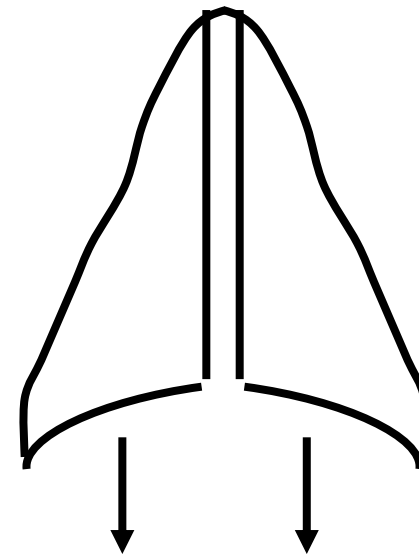
- Very weak muscles between the ribs
- Chest is very soft and flexible
- Diaphragm is primary muscle used to breathe

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

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



Make today a breakthrough.

Nutrition

- Rebecca Hurst Davis, MS, RD, CSP, CD
 - Intermountain Healthcare
 - becky.hurst-davis@imail.org

Objectives

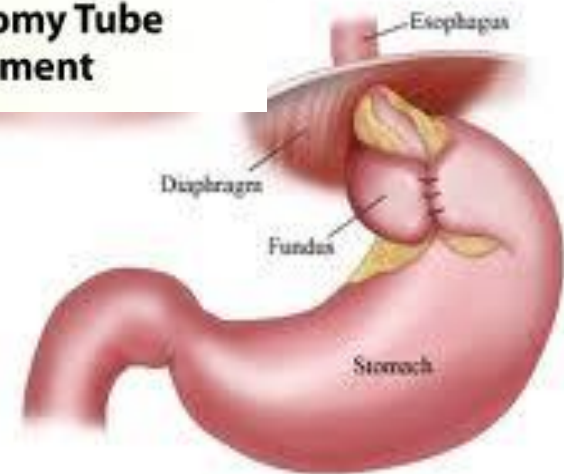
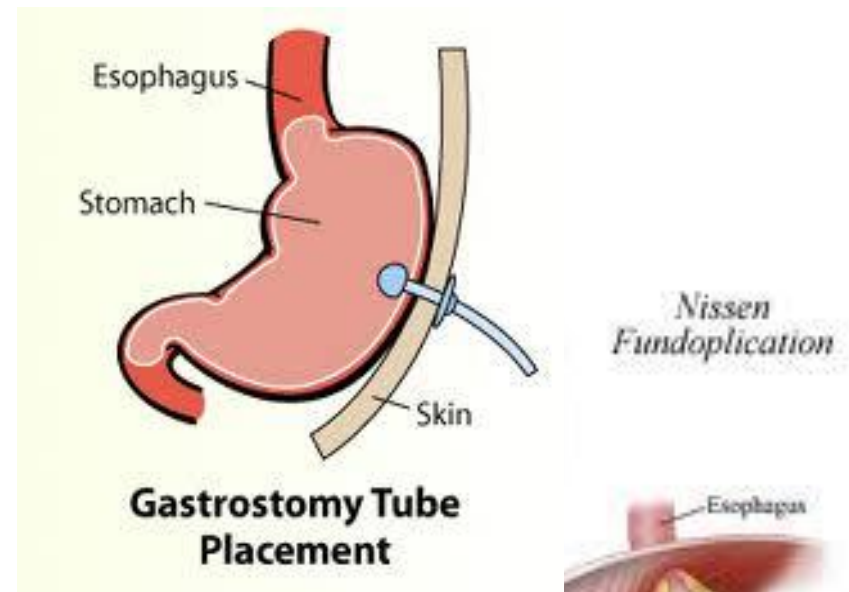
- Highlight the most important nutritional issues for children with Type I SMA
 - Safe Feeding/Feeding Tubes
 - Nutrition Referrals
 - Illness
 - Diet

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



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Options for Care...



- Vanessa Battista, RN, MS, CPNP
 - The Children’s Hospital of Philadelphia
 - Pediatric Advanced Care Team (PACT)
 - 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

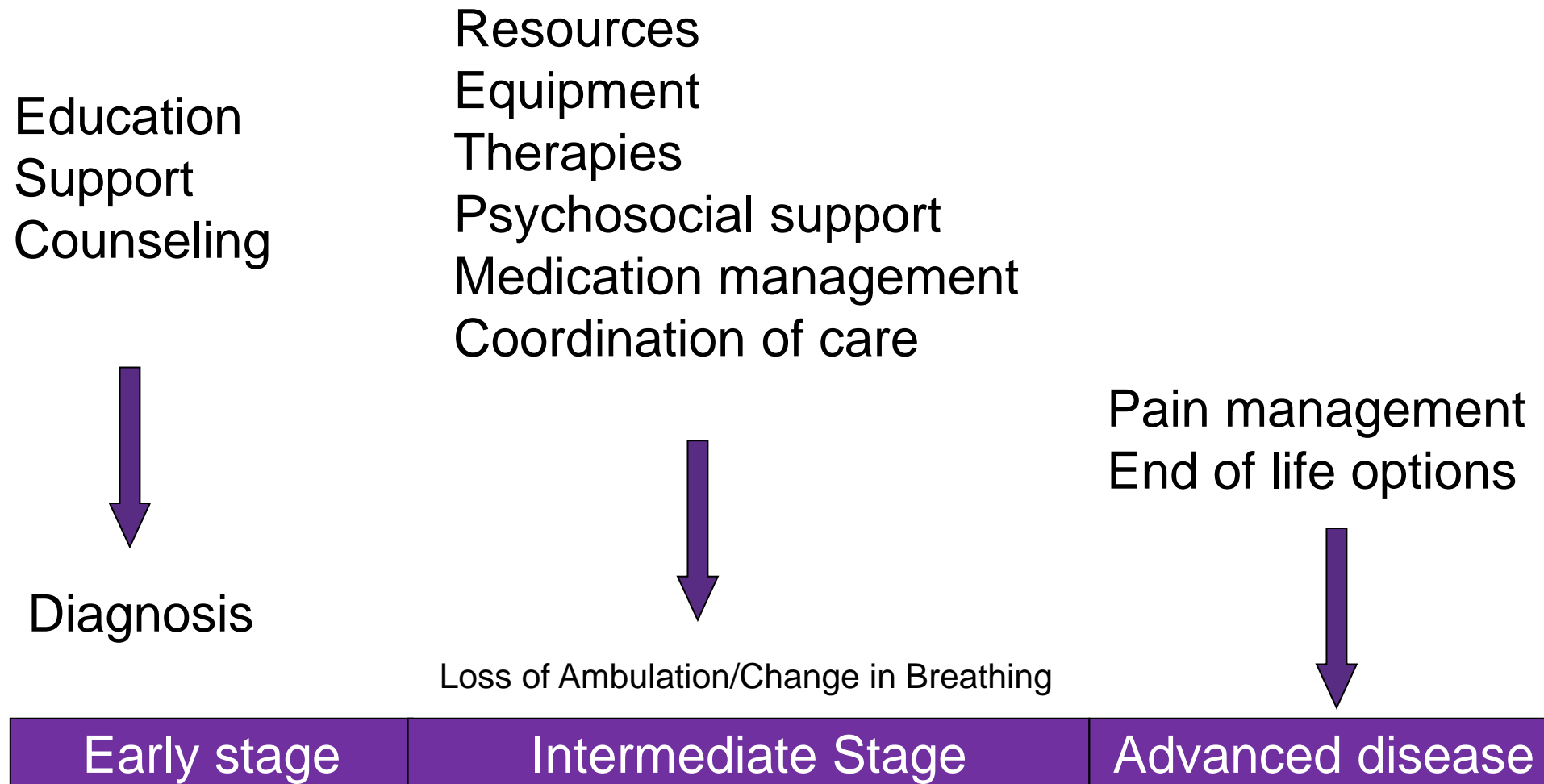
What Does Palliative Care Mean for My Family?

- 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
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 - 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



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- Karen Patterson, MS, PT, PCS
 - University of Wisconsin-Madison Doctor of Physical Therapy Program
 - Faculty Associate
 - CureSMA Medical Advisory Board
 - 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.**

