Palliative Care: When Should the Palliative Care Team Be Included in the Neuromuscular Team?

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

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Objectives for this Session

• Define pediatric palliative care

• Explore the relationship between pediatric palliative care (PPC) and Duchenne Muscular Dystrophy

• Explore families’ perspectives on decision-making regarding available care options for their children
What is Pediatric Palliative Care?

Why is it relevant or important in neuromuscular disease?
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
Palliative Care Goals

• Improve quality of life

• Provide collaborative & interdisciplinary care

• Support children’s & families’ goals/wishes

• Relief of pain and symptoms

• Attend to emotional, spiritual, and social needs

• Support for family and community members
Driving Principles

- Family-centered
- Specialized medical care
- An extra layer of support
- Living well until time of death
Palliative care is about aligning patients’ and families’ goals, hopes, and values with disease directed treatments.
Tenants of Decision-Making

Understanding goals of care
- Who is your child?
- What is most important regarding your child’s care?

Exploring options
- What are your hopes and fears regarding your child’s illness?
- What trade-offs are you willing to make for more time?

Using goals to help with decisions
- Where do you find support and strength?
- Is religion and/or spirituality important?
- Does your family make decisions privately or with others?
Integrative Model of Care

(Kobler et al., 2011)
A Snapshot of the PPC Patients in Hospitals

- Prospective observational cohort study
- 6 sites in the United States and Canada, with established hospital-based PPC teams
- Patients served by teams from January-March 2008 were eligible for enrollment
- Subjects were observed for 3 months
Results: Demographics

Age of Study Group n=512

- 10-18 years: 30.1%
- 19 years: 15.5%
- 1-9 years: 37.2%
- 1-11 months: 12.5%
- <1 month: 1.7%
Leading Conditions in Pediatric Palliative Care

- Genetic and congenital disorders: 41%
- Neuromuscular disorders: 39%
- Cancer: 10%
- Respiratory conditions: 7%
- Gastrointestinal: 8%
- Cardiovascular: 13%
- Metabolic: 20%
- Renal, immunologic: 10%

(Feudtner et al., 2011)
## Medical Technology of Pediatric Palliative Care Patients

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<thead>
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<th>Medical technology</th>
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<tr>
<td>Any feeding tubes</td>
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<td>Noninvasive ventilation</td>
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<tr>
<td>VP/VJ shunt</td>
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<td>2.9</td>
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(Feudtner et al., 200...
Signs and symptoms of patients who received pediatric palliative care consultation services:

(Feudtner et al., 2011)
The Goals of PPC in DMD

Support for the whole family throughout their life journey with NMD
Quality of Life

Physical
- Functional Ability
- Strength/Fatigue
- Sleep & Rest
- Nausea
- Appetite
- Constipation
- Pain

Psychological
- Anxiety
- Depression
- Enjoyment/Leisure
- Pain Distress
- Happiness
- Fear
- Cognition/Attention

Social
- Financial Burden
- Caregiver Burden
- Roles & Relationships
- Affection/Sexual Function
- Appearance

Spiritual
- Hope
- Suffering
- Meaning of Pain
- Religiosity
- Transcendence

Adapted from Ferrell, et al. 1991
Integrating Palliative Care in Life-Limiting Pediatric Neuromuscular Conditions: The Case of SMA-Type I and Duchenne Muscular Dystrophy

- Identified four transition points and opportunities for PPC at each stage
  - Peri-diagnostic
  - Treatment decision-making
  - Life-threatening events
  - End-of-life

Rushton, Erby, Cohn, & Geller, 2012
A Model for Neuromuscular Disease

Family-Centered Palliative Care for Neuromuscular Disease

- **Education**
- **Support**
- **Counseling**
- **Relationships**

- **Resources**
- **Equipment**
- **Therapies**
- **Psychological support**
- **Social support**
- **Medication management**
- **Coordination of care**

- **Pain management**
- **End of life options**

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**Diagnosis**
- Early stage

**Loss of Ambulation**
- Intermediate stage

**Change in Respiratory Status**
- Advanced disease
Interdisciplinary Team and Effort

- Neurologist
- Pulmonologist
- Palliative Care
- Cardiology
- PT/OT/Rehab Medicine
- Orthopedics/Neurosurgery
- Gastroenterologist (Nutritionist)
- Neuropsychologist
- Clinical Geneticist
- Primary Care Provider

IDT
Palliative Care Partnership

Interdisciplinary Team Members

Coordinate consultations
Neuromuscular Clinic

NP/Care Coordinator

Hospital Pediatric Advanced Care Team

Advisory Role

Ongoing exchange and feedback
What Are the Options?

Families should make choices that are consistent with their personal beliefs and values and which work best for them.
Goals of Care

• Essential to identify for the child and family

• Often relate to specific therapy or technology

• Fluid and continually changing

• Center upon (clinical) decision making
Respiratory Care

• Non-invasive
  e.g.- CoughAssist™ or BiPAP or a 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

Palliative Care

Less invasive
Changing textures of food

More invasive
Tube placed in stomach
Inherent Challenges

• **Communication**
  • family members
  • specialists from a variety of disciplines

• **Uniformity of Care**
  • duplicating or conflicting messages

• **Providing Care**
  • supporting families during stressful times
  • adapting care to ongoing changes
The relief of suffering and the cure of disease must be seen as twin obligations of a medical profession that is truly dedicated to the care of the sick.

Failure to understand the nature of suffering can result in medical intervention that (though technically adequate) not only fails to relieve suffering but becomes a source of suffering itself.

The nature of suffering and the goals of medicine, Cassel EJ NEJM-1982
Conclusion

• PPC provides care for a child and family along a physical, psychological, emotional, social and spiritual continuum.

• Children and families living with DMD can benefit from palliative care in a variety of ways.

• Various supportive care interventions exist and it is the role of PPC providers to offer information and support in navigating care options with children and families and the NMD team.
Acknowledgments

• Interdisciplinary NMD team members at Columbia University Medical Center, Children’s Hospital Boston, and The Children’s Hospital of Philadelphia

• Parent Project Muscular Dystrophy

• CHILDREN, ADOLESCENTS, AND FAMILIES LIVING WITH NMD
References


