

Palliative Care of the Patient with Neuromuscular Disease

Julie Hauer, MD + Michelle Meiring
UCT Paediatric Refresher Course
February 13, 2013

Dr Julie Hauer



A JOHNS HOPKINS PRESS HEALTH BOOK

Caring for Children Who Have

SEVERE NEUROLOGICAL IMPAIRMENT



A
Life
with
Grace



JULIE M. HAUER, M.D.

GREETINGS FROM BOSTON



What is the role of palliative care in DMD?



Palliative Care

- An approach to care that seeks to improve the **quality of life** of patients with life-threatening conditions and their families

WHO Definition of Palliative Care

Palliative Care

- **Aims to improve the quality of life** of patients and their families through early identification and treatment of pain and other problems, whether physical, psychosocial, or spiritual
- **Interdisciplinary** care for patients with serious, complex illness that addresses suffering and provides assistance with medical decision-making

Palliative Care

- Supports all treatment options
- Not just End-of-Life care
- Areas of support
 - Symptom treatment
 - Decision making
 - Respite care

Symptom management in NMD

- Pain
- Symptoms other than pain:
 - Constipation
 - Dyspnoea
 - Fatigue
 - Depression

Respite care



Helen House Oxford, United Kingdom



Douglas House

To provide care for young adults aged 16 – 35years



The Bar at Douglas House

Decision making

- Framework
- Interdisciplinary discussion
- Difficult discussions
- No right answer: The right choice is the one that is right for the individual child and family
- Resource constraints

Matt Johnston



<http://www.divingadream.org/>

Matt Johnston



In 2006, Matt ventured to the Florida Keys, where he became the first in his condition to experience diving in the ocean. The event was covered by NBC's Today Show.

Andrew

- Almost 18 year old with DMD
- Declining respiratory function and nutritional status
- Mild cognitive impairment
- Passive involvement, let's mom make decisions
- Did not share his wishes very openly
- Gave assent to his mom to limit interventions,

Andrew

- Later voiced his appreciation that his mom understood what he didn't want without having to say it
- He was able to later share that he didn't want increasing use of non-invasive respiratory support and he didn't want to be intubated;
- His main goal was to stay home, no more hospitalization, and to be comfortable.

Unique Themes

- Increasing dependence on others at a time that adolescents develop independence
- Transitioning from assent to consent

Introducing Concepts of Decision Making



Decision Making

- An opportunity to be informed, not a need for a decision
- How will the outcome look
- Level of evidence
- Conflicts or bias

Decision Making

- What are preferences and goals?
- How does the family approach decision making?
- How is information shared?
- Who should be present?
- What community resources exist to support a specific decision?

Goals and preferences may depend upon:

- Values
- Prior experiences with health care
- Prior experiences with death
- Family dynamics, coping skills, support
- Community resources
- Spiritual, religious, cultural, ethnic background and beliefs

Goals of Care

- Comfort
- Maintaining health without use of invasive interventions
- Medical care that can occur at home
- No hospitalizations
- Maintain health for upcoming family event

Advance Directive Discussion

- Prior to surgery:
 - Gastrostomy-tube
 - Spinal surgery
- Risk for life threatening event
- Decline in health or comfort
- Admission to the hospital

Try to avoid having to make decisions at the time of a crisis....

MOUNT SINAI JOURNAL OF MEDICINE 77:394–397, 2010

394

ETHICS

**Joseph's Wishes:
Ethical Decision-Making in
Duchenne Muscular Dystrophy**

Leigh Penner, LCSW,¹ Rabbi Mollie Cantor, BCC,¹ and Linda Siegel, MD²

¹Mount Sinai Kravis Children's Hospital, New York, NY,

²Department of Pediatrics, Albert Einstein College of Medicine, Bronx, NY

Whose decision is this?

- Explore understanding of situation
- Explore decisional capacity
- Rule out depression/suicidality
- Explore parental reasons
- Negotiate...

No matter what is decided...

- Continue to support the child and family
- Key philosophy of palliative care is “non-abandonment”
- Don’t say there is “nothing more that can be done”
- Support the terminal phase
- Meet families in bereavement

In conclusion...

“You matter because you are you. You matter until the end of your life

We will do all we can not only to help you die peacefully but to help you live until you die.”

Dame Cicely Saunders



You Betcha'

Be well

Do good work

And stay in touch

Garrison Keillor

Julie Hauer

julie.hauer@childrens.harvard.edu