Palliative Care of the Patient with Neuromuscular Disease

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Caring for Children Who Have Severe Neurological Impairment

A Life with Grace

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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 – 35 years

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/
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....
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

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