

PALLIATIVE CARE IN NEUROPEDIATRICS

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Nordic Neuropediatric
Conference

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Objectives

- Discuss problems due to severe impairment of the central nervous system that can remain intractable to treatment options
- Define tipping points that can prompt anticipatory discussions, including suggested recommendations that best meet goals
- Utilize language strategies that assist families when prognosis is not possible

Overview

- **Pediatric palliative care:** aims to relieve suffering, improve quality of life, and facilitate informed decision-making
- **This session:** the role of palliative care for children with severe impairment of the central nervous system (CNS)
- **Framework, Literature, and Data**

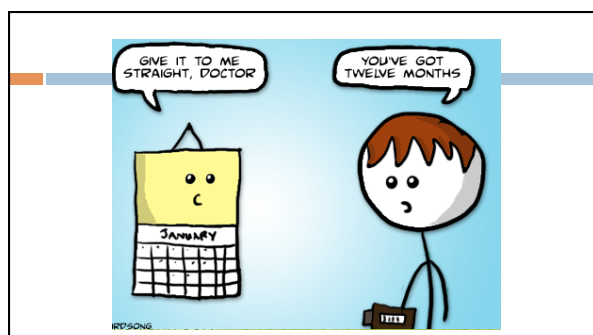
Barry

- 15 year old, intractable epilepsy, Bi-pap, inpatient every 1-2 months past 6 months, hospitalization with mechanical ventilation
- "Far from his baseline"
- Goals: comfort, quality of life, intubate if "reasonable" chance for recovery
- 3 hospitalizations for prolonged seizures

Courageous
Parents
Network:
videos with
his mother

Outcomes and Decision Making

- Prognosis: "how long will he live?", treatment outcome, recovery
- Prepare for various outcomes (manage uncertainty): families, teams, institutions
- **Lessen decisional regret**



Prognosis

Outcome based (death, development) and **Decision based** (surgery, DNI/ DNR)

Palliative Care

Process based (ability to define goals of care, allow hope)

Uncertainty to Certainty

Uncertainty of Outcome:

life span,
recovery,
benefit from
intervention



Certainty of Decision:

purpose,
intent, and
goal of a
decision

Bogetz, JF, Hauer J. 2018.

Hope

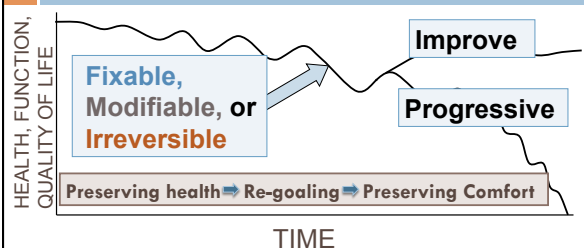
- **Adaptive:** to cope with adversity and loss
- **Affirmative:** to convey commitment and love



I've learned from my son that everything is possible, it doesn't mean everything will happen.

Feudtner 2009, Feudtner et al 2010

Hypothetical Trajectory



Intractable: not easily relieved or cured

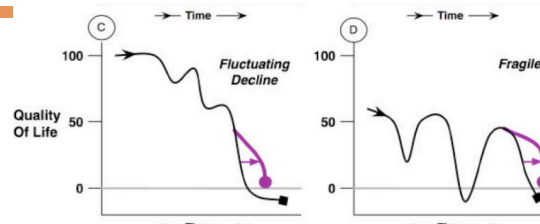
- Test and "Fix"
 - Fracture
 - Renal stones
 - Bladder infection
 - Respiratory infection
- Modify with risk for intractable
 - Seizures
 - Dysautonomia
 - GI motility
 - Central pain
 - Mucus clearance

Quality of Life (QOL) Considerations

- QOL as indicated by parents of 14 children with profound intellectual and motor disability (PIMD)
 - ▣ Ability to enjoy
 - ▣ Comfort
 - ▣ Awareness
 - ▣ Multiple health problems
 - ▣ Falling below the lower threshold of QOL

Zaal-Schuller IH, et al. Considering quality of life in end-of-life decisions for severely disabled children. *Res Dev Disabil.* 2018;73:67-75.

Quality of Life (QOL) Considerations



Feudtner C. Collaborative communication in pediatric palliative care: a foundation for problem-solving and decision-making. *Pediatr Clin North Am.* 2007;54(5):583-607, ix.

End of Life Decision Making (EOLD)

- **Parents:** half indicated missed opportunity by physicians to discuss EOLD at time of stability
- **Physicians:** found it difficult to identify when a parent was “ready” for discussion
- **Physician discussion:** typically initiated at time of acute deterioration

Zaal-Schuller IH, et al. How parents and physicians experience end-of-life decision-making for children with profound intellectual and multiple disabilities. *Res Dev Disabil.* 2016;59:283-293.

Patterns of End of Life Care

- **Progressive** central nervous system disorders **versus**
- **Static** encephalopathy, congenital, genetic
 - Second group less likely to have palliative care consult, die at home, plan location of death, or have DNR orders in place
 - More likely to receive intensive life sustaining therapies in last 2 days of life

DeCoursey DD, et al. 2018.

	CNS-Progressive N=27	CNS-Static N=19
Palliative care consult	24 (89)	10 (53)
Any DNR order	20 (74)	8 (42)
Death: ICU/Hosp/ED	12 (44)	12 (63)
Death: Home or Other	15 (56)	7 (37)
Comfort at EOL	18 (67)	4 (21)
Vent and/or CPR	1 (4)	4 (21)
Plan Location	16 (59)	9 (47)

DeCoursey 2018, Values are N (%), ED=emergency dept, EOL=end of life

Seven Hills Pediatric Center

- **Criteria for Respite or Long Term Care:**
 - ▣ Development <12 months
 - ▣ 2 or more complex care needs
 - ▣ Admit prior to 22 years, remain life long
- **Reasons to use Long Term Care**
- **Medical care comparable to home**

34 deaths: general characteristics

- **Average age:** 22 years
- **Age range:**
 - ▣ 11 months to 50 years
 - ▣ 10 (29%) older than 30 years
- **Conditions:** anoxic and traumatic brain injury, neuro-structural malformations, in-utero infection, genetic, metabolic

Hauer J, et al. Anticipating Death in Children and Adults with Childhood Onset Severe Central Nervous System Impairment: A Case Series Review (FR480). *J Pain Symptom Manage*. 2018;55(2):631.

General Characteristics

Feature / Problem	N=34
Feeding tubes	N=34 (100%)
Anti-seizure drugs	N=33 (97%)
Complex symptom management	N=23 (68%)
Respiratory: chronic, recurrent	N=15 (44%)
Gastrointestinal: acute recurrent	N=10 (29%)
Ventilation: invasive, noninvasive	N=7 (21%)

Goals Discussion N=27 (79%)

- **When:** symptom burden, decline, decreased engagement in activities, recurrent ileus / intestinal pseudo-obstruction, EOL features
- **Identifying burden, risk, medical reason**
- **Proactive and reflective**
- **Meeting with team and parents**

Goals Discussion N=27 (79%)

- **Common goals:**
 - ▣ To be comfortable, To not suffer
 - ▣ To get better, To live as long as possible
 - ▣ To be with family (including “extended”)
- **Missed opportunity in 4 others**

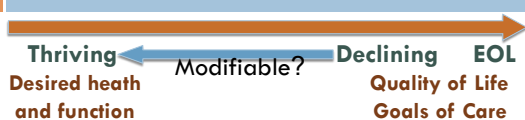
Zaal-Schuller IH, et al 2018. Tamburro RF, et al. 2011

Redirection of Care in 23 of the 27

Redirection of care to death: hours to 5 years

Reasons for Redirection	N= 23 of 34
General decline	N=19 (55%)
Global symptom burden	N=16 (47%)
End of life (EOL) features	N=16 (47%)
Respiratory: chronic, recurrent	N=15 (44%)

Quality of Life and Goals of Care



Continued decline likely when 2 or more categories identified: Symptoms, QOL, intractable problems, EOL features

End of Life (EOL) Features N=16 (47%)

- Features at EOL in the last hours / weeks
- Features:
 - Ileus and/or edema without cause N=13
 - Change in respiratory pattern
 - Remain unarousable
 - Changes in extremity color and temp to touch
- Preparing families and staff for timeline

Ileus or Edema at End of Life

- Ileus and/or edema without cause: 13 (38%)
 - Acute or recurrent ileus: N=10 (30%)
 - Edema: N=6 (18%)
- Assessment: metabolic panel, urine analysis and culture
 - **11: pre-existing decline in health, function, QOL**
 - 2 with severe anoxic brain injury
- Management: review options, provide suggestions

Siden H, et al. Pediatric enteral feeding intolerance: a new prognosticator for children with life-limiting illness?
Hauer J. Feeding Intolerance in Children with Severe Impairment of the Central Nervous System

Language Suggestions

- “These features worry me...”
- “The tests were normal. It is unlikely that there is a fixable reason for his intestines shutting down. This can be due to the body shutting down or changes in the area of the brain that regulates the gut. In some, gut function won’t improve.”

Hauer J. Feeding Intolerance in Children with Severe Impairment of the Central Nervous System: Treatment and Prevention. Children (Basel). 2017; 5(1). pii: E1.

Language Suggestions

- “We will give an amount of fluid by G-tube that his body is more likely to tolerate. We will discuss what that means if he doesn’t...”
- “I’m glad we are protecting him from feeds/ fluids that his body can’t process at this time”
- Decision to hold feeds/fluids: no regret, “lesser of 2 evils”, “the only thing that made any sense”

Rapoport A, et al. Pediatrics. 2013;131(5):861-9.

Language Suggestions

- **Acknowledge the emotion:** I see your distress, I am so sorry for how hard this is for you.
- **Address the concern:** Holding feeds and decreasing fluids has lessened the swelling and fluid in his lungs that was causing him so much discomfort.
- **Redirect to an achievable goal:** His face looks so relaxed, do you have the same observation?

Respiratory Features

- Respiratory distress: active respiratory effort and unmet symptom burden, uncommon at EOL
- Opioid use in 15:
 - 13 (86%) on scheduled morphine: months to years
 - "I think he lived longer because of this"
- Acute and chronic treatment
- Change in respiratory rate and rhythm at EOL:
 - apneic pauses, Cheyne-Stokes, agonal

Chronic Ventilation N=7 (21%)

- Trach with ventilation N=4
- Non-invasive ventilation N=3
- Discontinued in 5 to meet goals of care, due to changes in QOL

Goals of Care and Decision Making

- Altered QOL (alertness, comfort)
- Hopes/Goals: improved health, alertness, and comfort
- "I've heard you mention several hopes, let's review what might be possible"

Goals of Care and Decision Making

- Follow-up: “I wish...” “I wonder...”
- Discussion when technology not offering the benefit it once did
- “I’m glad for all the years of benefit...”
- Introduced with no need for decision

EOL care at SHPC, N=25 (74%)

- Found without pulse N=3
- Active adjustments in care plan N=22
 - ▣ Symptom management
 - ▣ Adjust feeds / fluids at end of life
 - ▣ Stop nonessential interventions
- Requires experts in EOL care

Deaths Away from Facility N=9 (26%)

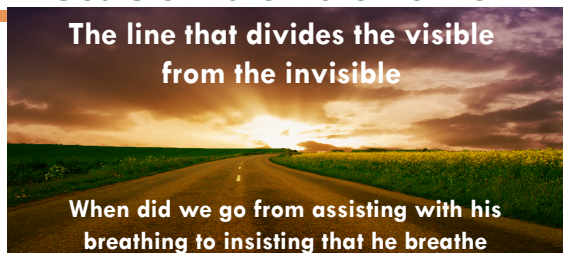
- Resuscitation in emergency room N=4 (12%)
- Decision to withhold/withdraw N=5 (15%)
 - Trial of ventilation N=2
 - Following GI surgery N=1

What can we “measure”

- Measure the right things, set the right targets
- ▣ Trajectory, Quality of Life
 - ▣ Ability to further modify outcome
 - ▣ Ability of care plan to meet goals
 - ▣ Altered CNS function
 - ▣ Reassess as needed

Goals of Care – the Horizon

The line that divides the visible
from the invisible



When did we go from assisting with his
breathing to insisting that he breathe

Communication Skills

- “I wonder...”
- “I wish... but at this stage it is not possible...”
- “I heard you mention several hopes, at this stage we can’t make that first one happen”
- “Loving parents...”

Feudtner C. Collaborative communication in pediatric palliative care: a foundation for problem-solving and decision-making. *Pediatr Clin North Am.* 2007;54(5):583-607, ix.

Barry: 15 year old, intractable seizures

- “I wish he could be seizure free, I wonder...”
- **What we can do:** “I will review treatment options”
- **What we may not want to do:** “It might make sense not to use (intubation, IVs) given that it doesn’t improve his (seizures, GI), what are your thoughts...?”
- **Offering options:**
 - Location of Care
 - Non-invasive management (home or hospital)
 - Use at home, Trial in the hospital

Barry: 15 year old, intractable seizures

- 6 months later: Bipap discontinued
- 1 ½ years later: acute ileus: “One option is to send him to the hospital to place an IV. I think the best plan is to use a trial of pedialyte by feeding tube.”

Conclusions and considerations

- Proactively monitor for significant decline in quality of life and burden of treatment
- Determine Goals of Care at such times and revisit as needed
- Offer treatment plans that best meet these specific goals

Conclusions and considerations

- Comfort at End of Life (EOL): decisions to not use invasive interventions at “tipping points” including acute ileus or edema in the context of decline
- Majority of deaths: occurred following decision to forgo or discontinue life sustaining therapy
- Option of hospital for noninvasive management
- EOL care requires expertise

Thank you!

Keep in touch!

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