Pediatric Palliative Care in the 21st Century: Live as long and as well as possible

Stefan J. Friedrichsdorf, MD, FAAP
Medical Director, Department of Pain Medicine, Palliative Care & Integrative Medicine
Children's Hospitals and Clinics of Minnesota, Minneapolis/St. Paul, MN

Learning Objectives

- Review common obstacles for referral to Pediatric Palliative Care (PPC)
- Evaluate top myths in PPC
- Underscore importance of interdisciplinary team approach in PPC
# Palliative Care

- **Palliare (Latin):** to cloak
- Palliative care is about matching treatment to patient goals.
- Specialized medical care for children with serious illness
- Focused on relieving pain, distressing symptoms & stress of a serious illness
- Appropriate at any age and at any stage, together with curative treatment
- Goal is to improve quality of life for child/family

## Myths, Misconceptions, and Assumptions...

- You are taking care of a seriously ill child. You would not be surprised if she might die within the next few months. You are considering a palliative care consult...
- What are arguments might you hear from colleagues (or family) not to do that?

## Assumption #1:

**The death of a child is a rare event** (especially in the USA...)
Death of Children

- 20 children die worldwide every second
- 1,200 children die worldwide every hour
- 29,000 children die worldwide every day
- 15-20 million children would benefit from PPC annually (low-estimate)
- ... and in the US!

Causes of Death in Children 0-19 years (USA, 2013)

1. Infant Mortality (<1 year) 23,440
   Fetal Deaths > 20 wks gestation 24,073
2. 1-19 Years 18,888
3. Life-limiting diseases >10,800
   Congenital malformations, chromosomal abnormalities 5,740
   Malignant Neoplasms 1850
   Heart Disease 493
4. Accidents 7,645
5. Homicide 2,021
6. Suicide 2,143

Total 42,328

More than 115 children die in the US every day…

- More than 1 child every 15 minutes…
Life-Limiting Conditions (LLC)

- ...are those for which there is no reasonable hope of cure and from which children will die before reaching adulthood.

  - White: 37/10,000
  - Chinese: 32/10,000
  - Black: 42/10,000
  - South Asian: 48/10,000

  - Prevalence [32/10,000]: > 237,000 with LLC
  - Mortality [1.5-1.9/10,000]: 10,800 - 13,700 die/year ACT & Royal College of Paediatrics and Child Health, 2003
  - 15,000 die/year Age (0-24), who would benefit from PPC Feudtner, 2001

Boeing 747-400
416 passengers

US Health Care System...

- USA: (conservatively estimated)
- 237,000 children live with life-limiting conditions (LLC)
- 570 "Boeing 747"
- 10,800 - 13,800 children 0-17 years die each year due to life-limiting conditions
- 26-33 "Boeing 747"
  - one crash every 11-14 days
Assumption # 2:

Pediatric Palliative Care is usually for children with cancer...

Causes of death in children due to life-limiting conditions


Pediatric Cancer: Epidemiology USA

- New cancer diagnosis: > 16,000 children (0-19 years) / year
- Large majority are cured of their malignancy (> 80% of children with cancer are alive 5 years after diagnosis [62% in the mid-1970s]).
- However, children with good fortune to attain cure nonetheless experience considerable suffering
- 1,960 children and adolescents are expected to die due to a malignancy in 2014
Pediatric Cancer Survivors


Access to PPC?

- Comprehensive palliative care is the expected standard of care for patients with advanced cancer, however access to, and availability of palliative care expertise for the majority of children with life-threatening conditions, is still lacking compared with adult services.

- In the U.S., the vast majority of infants, children, and teenagers with advanced illnesses who are near the end of life do not have access to interdisciplinary pediatric palliative care (PPC) services either in their community or at the nearest children's hospital.

Sibling Care

“**What do you gotta do to get some attention in this family...?”**


- 99% of siblings present at time of death NOT regretting being there; 76% of siblings NOT being present, regretted it. Lovgren, M., et al., Siblings' experiences of their brother's or sister's cancer death: a nationwide follow-up 2-9 years later. Psychooncology, 2015.

- PPC referral plus positive family functioning = normal psychological
Assumption # 3:

The “sudden death”...
“Sudden” Death? Advanced Illness Marked by Slow Decline with Periodic Crises and “Sudden” Death

Multiply relapsed cancer
Cystic Fibrosis
Advanced HIV
Refractory Seizure Disorder
Solid organ transplant recipient
Congenital heart disease

Assumption #4:

Specifically trained Pediatric Palliative Care specialist are not required...
### Fatigue 97%
3% 18%

### Pain 82%
28% 76%

### Dyspnea 82%
17% 64%

### Poor appetite 82%
4% 38%

### Nausea and vomiting 58%
10% 42%

### Constipation 51%
10% 42%

### Diarrhea 42%
1% 18%

---

#### An early palliative care intervention (even from the point of diagnosis) = appropriate and beneficial treatments, increased quality of life and may in fact lead to prolonged (I) life.

- RCT, n=151; adult cancer patients receiving palliative care early in their illness lived longer (11.6 months vs. 8.9 months, P<0.02), with better quality of life, including decreased depression
- This difference was despite less aggressive end-of-life care
- Results underscore the need for palliative care early in a serious illness
- This appear to refute the notion that palliative care means giving up. Patients received palliative care alongside their curative treatment.
PPC Prolongs Survival

- Kids: two cases of Herlitz-type EB, where early palliative care (PC) improved pain and symptom management so dramatically that the patient with PC involvement lived significantly longer than the patient with later PC involvement.


- Adults: In ENABLE III study, patients who received early palliative care had significantly longer 1-year survival rates than those who received delayed palliative care; the difference was 15% at 1 year (63% vs. 48%; p = .038).


- Chemotherapy for end-stage cancer does not prolong life, but reduces quality of life. (n=158, 3.8 months before death)


Assumption # 5:

Pediatric palliative care starts:
- when curative treatment stops,
- and when a child is close to dying
- and ends at death
Pediatric Palliative Care

"Palliative care no longer means helping children die well, it means helping children and their families to live well and then, when the time is certain, to help them die gently."

Mattie Stepanek, 1990-2007

Pediatric Palliative Care

- Earlier recognition by both physicians & parents that child had no realistic chance of cure led to stronger emphasis on treatment to lessen suffering & integrate PPC in pediatric cancer patients

- Religion, Spirituality or Life Philosophy play an important role in live of most parents whose children receiving PPC
  - FICA Spiritual Assessment Tool (Puchalski 2010)

- Focused on relieving pain, distressing symptoms & stress of a serious illness

- Goal is to improve quality of life for child/family

- Link between spiritual coping and outcome for adolescents
Curative Care? Or Pediatric Palliative Care? Well...the answer is yes!

Assumption # 6:

- Parents have to choose between “Fighting For a Cure” or “To Give Up”
- PPC translates into “Giving Up Hope” and “Doing Nothing”

...in face of serious illness
Hope in face of serious illness

- Notably, 40% of clinicians believe that caring for patients with poor prognoses is depressing, and this was more common among less-experienced clinicians.

Continued treatment in face of serious illness

- In discussions of treatment options with families, Wolfe and Grier suggest "The very nature of miracles is that they are rare. However, we have seen miracles, and they have occurred both on and off treatment." Pizzo, P.A., Poplack, D.G. (Eds) (2002) Principles and Practice of Pediatric Oncology (4th edn). Philadelphia, PA: Lippincott Williams & Wilkins.
- Motivated either by hope for a miracle, desire to extend life, or desire to palliate symptoms related to progressive disease.

- In other words, a child does not have to continue on disease-directed therapy in order to preserve hope, especially when the therapy significantly impacts child's remaining quality of life.
- Regardless, decisions regarding continued disease-directed therapy need to be carefully considered, weighing the potential for life extension and impact on quality of life.
- Even when the underlying condition can not be cured, sophisticated medical technology will be used to control symptoms and improve a child quality of life.
- It is a very active and advanced approach to pain & symptom management and family support.
Hope

- Caring for a dying child is emotionally very difficult.
- It may be particularly challenging for physicians and other caregivers to consider the integration of palliative care because this may be perceived as 'giving up'.
- More importantly, parental loss of a child is certainly considered to be the most difficult type of loss.

- As a result, the emotional cost of recognizing that a child may die impedes planning for optimal care and support.

Chew-Sheet Hope Language 1/2

- Tell me about little Claire on a good day! Do you have any pictures?
- Considering what little Johnny is up against, what are you hoping for?
- I am hoping for a miracle, too. And I have seen miracles, but they are very rare and happen on treatment or off treatment...
- Just in case, the miracle or the cure is not going to happen (...if God/Allah has different plans for Sarah), what else are you hoping for?
- We want to make sure that Karen lives as long as possible, as well as possible!
- We are hoping for the best, but preparing for the worst.
- “He is not dying because he is not eating...he is not eating, because he is dying...”
Cheat-Sheet Hope Language 2/2

- "Loving parents…" (…equally loving parent realize that the child has come to the natural end of their life…)
  - "I wish…, but at this stage it is not possible…"
  - Responding to emotion "I can’t imagine what this might be like for you"
  - "I am aware of the time…"
- So what I hear you saying, is the following...
  - Did I get this right…!
- Then I would recommend the following...
- DNR/DNI (AND, limiting of painful interventions)
- I recommend to put in an order to protect your daughter from experiencing painful situations, such as chest compressions or intubation, in case the breathing or heart stops - are you fine with that?

Assumption # 7:

Increasing the dose of opioids causes respiratory depression and quickens death

(Morphine = Euthanasia?)

Opioids for Pain & Dyspnea

- "Morphine kills the pain, not the patient" MD killing patient in name of pain relief is not merciful, just incompetent, just incompetent Sykes N.P., Morphine kills the pain, not the patient Lancet. 2007; 370:921-925.
- An enduring misconception is the belief that in the management of pain and dyspnea, opioids will hasten death and should only be administered as a last resort. This was contradicted in the adult literature… Thormann T, Sykes N. Opioid use in the last week of life and implications for end-of-life decision making Lancet. 2002;332:298-304.
- …and our PPC team commonly observes that administering opioids and/or benzodiazepines, together with comfort care to relieve dyspnea and pain, not only prolongs life but also improves the child’s quality of life. Thorns A, Sykes N. Opioid use in last week of life and implications for end-of-life decision making. Pediatric Hematology Oncology. 2000;17:398-400.
- Retrospective cohort study (n=223 adult oncologic patients)...Pain management in children with advanced cancer and during course of life care. Pediatric Hospice Care 2010 May 27(5):357-64.
  - Mean Survival:
    - < 2-fold increase in their initial opioid dose = 9 days
    - > 2-fold increase in their initial opioid dose = 22 days

• "Morphine kills the pain, not the patient" MD killing patient in name of pain relief is not merciful, just incompetent Sykes N.P., Morphine kills the pain, not the patient Lancet. 2007; 370:921-925.
• An enduring misconception is the belief that in the management of pain and dyspnea, opioids will hasten death and should only be administered as a last resort. This was contradicted in the adult literature… Thormann T, Sykes N. Opioid use in the last week of life and implications for end-of-life decision making Lancet. 2002;332:298-304.
• "Morphine kills the pain, not the patient" MK killing patient in name of pain relief is not merciful, just incompetent. Thormann T, Sykes N. Opioid use in the last week of life and implications for end-of-life decision making Lancet. 2002;332:298-304.
• An enduring misconception is the belief that in the management of pain and dyspnea, opioids will hasten death and should only be administered as a last resort. This was contradicted in the adult literature… Thormann T, Sykes N. Opioid use in the last week of life and implications for end-of-life decision making Lancet. 2002;332:298-304.
Assumption # 8:

PPC takes patients away from primary care / pediatric specialists

Primary versus Subspecialty PPC

- Why is it, that we ask an infectious disease specialist for a consult, if we can prescribe antibiotics ourselves?

  **Primary PPC**
  - Team provides interdisciplinary support and integrates a palliative care approach

  **Subspecialty PPC**
  - Clinical: more complex care

- Skills in advanced pain & symptom management
- Education: enhance knowledge, skills, attitudes & behaviors
- Innovation and research: advancing the field
- Advocacy: system wide changes
Pediatric Palliative Care Consult

- Complementary
- “How can we help...?”
- May involve second opinion regarding
  - Decision making
  - Symptom management
  - Coordination of care
  - Home care

Assumption # 9:

A part time (0.2-0.5 FTE) physician alone can certainly address all pertinent PPC issues in a Children’s Hospital

PPC

- PPC applies to all children with life-threatening illness
- PPC Specialists provide:
  - Direct patient consultation
  - Advocacy
  - Education
  - Quality Improvement
  - Research
**PPC Example...**

- “If you have seen one Pediatric Palliative Care program ...
- ...you have seen one Pediatric Palliative Care program!

---

**The Business Case**

- 60% reimbursed fee for service
- Philanthropy supported 40% of program expenses in 2014

---

**Why does Children’s fund a program “losing money”?**

- ...well, we do, but we don’t really...
### Pain & Palliative Care: Reduce staff turn over

- 30-50% of all new RN’s elect either to change positions or leaving nursing completely within the first 3 years of clinical practice (ANCC, 2003; Airken, Clarke, Sloane, Sockalski, & Silber, 2002; Cipriano, 2006; Cowin & Hengstberger-Sims, 2006).


- Reducing turnover and improving overall job satisfaction and performance important in assuring patient and family satisfaction while promoting quality care.

### Hospital Support

- Recognition
- Increases staff efficiency
- Halo Effect: Increase in market share
- Decrease in complications / earlier discharge = higher patient turn over
- Reduced liability for iatrogenic harm
- Increased philanthropy

- Increased staff / patient / parent satisfaction
- HCAHPS (Hospital Consumer Assessment of Healthcare Providers and Systems) survey
- Reduced staff turn-over (> $ 60-100K training 1 ICU RN)
- “Never Events” NOT reimbursed (PPC patients over represented) --> home care
- Decreased 30-day readmission

### Pediatric Palliative Care: Better Care & Cost Savings?


- Compare pediatric hospital resource utilization before and after enrollment
- Non-cancer patients: LOS decrease 38 days, decrease hospital charges $ 275,000 / patient
Pediatric Palliative Care is...

- Specialized medical care for children with serious illness
- Focused on relieving pain, distressing symptoms & stress of a serious illness
- Appropriate at any age and at any stage, together with curative treatment
- Goal is to improve quality of life for child/family
- Provided by an interdisciplinary team who work with the patient's other physicians & health care providers; provides an extra layer of care
- P.S.: Morphine & Midazolam do not shorten a child's life

Hope and PPC include each other

Pediatric Palliative Care:

- Multi--> Inter--> Trans-disciplinary Team

- (1) “How can we help?”
- (2) Then listen...

"If you can't add life to my son's days, then don't add days to my son's life"

Father of a toddler to Dr. Sunny Anand on PICU
Further Links


- Video: Kiran Stordalen and Horst Rechelbacher Pediatric Pain, Palliative and Integrative Medicine Clinic Tour [https://vimeo.com/124644881]


- Short Movie: Meet the Interdisciplinary Chronic Pain Clinic Team at Children’s Minnesota: Link2Sci TV [https://www.youtube.com/watch?v=i13kyc70844]

- Video: Tour of the Kiran Stordalen and Horst Rechelbacher Pediatric Pain, Palliative and Integrative Medicine Clinic at Children’s Hospitals and Clinics of Minnesota and an overview of the three programs that are offered at Children’s under this clinic [https://vimeo.com/133357296]

- Short Movie: LittleStarsFilm Kali’s Story - Beyond the NICU: This amazing pediatric palliative care short movie (7 min) features 8-year-old Kali’s journey at Children’s Hospitals and Clinics of Minnesota from NICU to today receiving care by the Pain & Palliative & Integrative Medicine program while inpatient, in the clinic, and at home (Jan 22, 2015) [http://www.littlestars.tv/littletv/beyond-the-nicu/]

Further Training

- 10th Annual Pediatric Pain Master Class
  - Minneapolis, Minnesota, USA | June 17-23, 2017

- Education in Palliative & End-of-life Care (EPEC): Become an EPEC-Pediatrics Trainer
  - Montréal, Québec, Canada | April 29-30, 2017 (Professional Development Workshop: 04/28/17)

Contact: CIPPC@ChildrensMN.org

Blog: http://NoNeedlessPain.org