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

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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)**  
   - Fetal Deaths > 20 wks gestation: 24,073
   - Total: 23,440

2. **1-19 Years**  
   - Total: 18,888

3. **Life-limiting diseases**  
   - Congenital malformations, chromosomal abnormalities: 5,740
   - Malignant Neoplasms: 1,850
   - Heart Disease: 693
   - Accidents: 7,645
   - Homicide: 2,021
   - Suicide: 2,143
   - Total: >10,800

Total: 42,328

http://www.cdc.gov/nchs/data_access/Vitalstatsonline.htm#Fetal_Death
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: 27/10,000
  - Chinese: 32/10,000
  - Black: 42/10,000
  - South Asian: 48/10,000

- USA - Age 0-17: 74.3 million children (2014) [http://www.childstats.gov/americaschildren/tables/pop1.asp]
  - 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]

How Many Children Would Benefit from PPC?

- UK: 32/10,000 (high income country)
- South Africa 150/10,000 (upper middle income country)
- Zimbabwe 180/10,000 (low income country)
- No accurate figure globally: ICPCN estimates 15-20 million children worldwide (low estimate)

- World Health Assembly Resolution 67.19 on Palliative Care [May 2014]: ensure equitable access to palliative care, including pain relief, for neonates, children and young people and their families

[http://www.icpcn.org](http://www.icpcn.org)
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

Germany: 0-15 years
- Cancer (31%)
- Neuromuscular/neurodegenerative (20%)
- Congenital or genetic (16%)
- Cardiovascular (12%)
- Metabolic (9%)

USA: 0-24 years
- Cancer (12%)
- Neuromuscular/neurodegenerative (20%)
- Congenital or genetic (16%)
- Cardiovascular (12%)
- Metabolic (9%)

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

- Prevalence of pain during treatment: Outpatients 9-26%, inpatients 39-54%
- Survivors: Prevalence of pain conditions (12% pain/abnormal sensation, 15.5% migraines, 20.5% other headaches) and using prescription analgesics higher among survivors than siblings.
- Pediatric brain tumor survivors experience many symptoms after treatment. Lack of energy (52%), difficulty with sleep (38%), lack of concentration (36%), and headaches (36%).

Pediatric Cancer: Epidemiology USA

- Germany: 0-15 years
- Germany: 0-15 years
- USA: 0-24 years

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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 been there; 76% of siblings NOT being present, regretted it Langemo, M. et al. Siblings’ experiences of their brother’s or sister’s cancer death: a nationwide follow-up 3-9 years later. Psychonomics, 2015.


http://opinionator.blogs.nytimes.com/2015/12/16/when-a-baby-dies
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

Outcomes Improved with PPC Involvement

- Children who received PPC/Oncology more likely to have fun (70% versus 45%) and to experience events that added meaning to life (89% versus 63%). Friedrichsdorf SJ et al. J Palliat Med 2015
Assumption # 4:

Specifically trained Pediatric Palliative Care specialist are not required...

103 Children With Cancer: Last Month of Life

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Prevalence</th>
<th>Distress</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fatigue</td>
<td>97%</td>
<td>18%</td>
</tr>
<tr>
<td>Pain</td>
<td>82%</td>
<td>76%</td>
</tr>
<tr>
<td>Dyspnea</td>
<td>82%</td>
<td>64%</td>
</tr>
<tr>
<td>Poor appetite</td>
<td>82%</td>
<td>38%</td>
</tr>
<tr>
<td>Nausea and vomiting</td>
<td>58%</td>
<td>42%</td>
</tr>
<tr>
<td>Constipation</td>
<td>51%</td>
<td>10%</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>42%</td>
<td>1%</td>
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</tbody>
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Symptom prevalence and distress reported by children with advanced cancer

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 (!) 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
- 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.

* Although this is only one study, it is an exciting one & results are not surprising: PC clinicians regularly see these outcomes in practice - even in pediatric patients. Temel JS, Greer JA, Muzikansky A, Katznelson S, Assia S, Jackson VA, et al. Early palliative care for patients with metastatic non-small-cell lung cancer. N Engl J Med. 2010 Aug 19;363(8):733-42.

Assumption # 5:

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

"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.
  


- FICA Spiritual Assessment Tool (Puchalski 2010)

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


- Most clinicians (90.3%) believed they were prepared to elicit parent's hopes (however only 21.2% patient's hopes). 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


- Motivated either by hope for a miracle, desire to extend life, or desire to palliate symptoms related to progressive disease.

- 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.
Continued treatment in face of serious illness

- 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.

Cheat-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

- 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 (Sykes, N.P. Morphine kills the pain, not the patient. Lancet, 2007;369(9570): p. 1224-6).
- 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... Thorns A, Sykes N. Opioid use in last week of life and implications for end-of-life decision making. Lancet. 2000;356:398-399.
  - Mean Survival:
    - < 2-fold increase in their initial opioid dose = 9 days
    - > 2-fold increase in their initial opioid dose = 22 days
Assumption # 8:

PPC takes patients away from primary care / pediatric specialists

<table>
<thead>
<tr>
<th>Primary versus Subspecialty PPC</th>
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<tbody>
<tr>
<td><strong>Primary PPC</strong></td>
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<tr>
<td>- Team provides interdisciplinary support and integrates a palliative care approach</td>
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<tr>
<td><strong>Subspecialty PPC</strong></td>
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<tr>
<td>- Clinical: more complex care</td>
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<tr>
<td>- Skills in advanced pain &amp; symptom management</td>
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<tr>
<td>- Education: enhance knowledge, skills, attitudes &amp; behaviors</td>
</tr>
<tr>
<td>- Innovation and research: advancing the field</td>
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<tr>
<td>- Advocacy: system wide changes</td>
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</tbody>
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**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 FTE) physician
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!
Conclusions

Not providing PPC to children with serious illness is now considered “bad”

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