Disclosures

- I have no relevant financial relationships or conflicts of interest to disclose.

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Overview

1) Understand the potential benefit from early palliative care consultation
2) Understand the value of concurrent palliative care with curative therapy
3) Understand the importance of effective communication in palliative care
4) Understand the principles of effective symptom management
5) Understand some of the Bioethical Issues in Pediatric Palliative Care

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#ChooseKnowledge
Epidemiology

- Approximately 500,000 children cope with life-threatening conditions annually in the United States.
- Approximately 20,000 infants and children die annually in the United States alone.


#ChooseKnowledge

Mortality rates for three US pediatric cohorts 1950 to 2016


#ChooseKnowledge

Epidemiology

Traditional Model of Medical Care

Disease-modifying therapy
(Curative, Restorative)

Palliative Care

Presentation of life-limiting illness

Death & Bereavement

National Hospice Work Group. Adapted from work of the Canadian Palliative Care Association & Frank Ferris, MD

NATIONAL CONSENSUS PROJECT

According to more recent teaching as set forth by the National Palliative Care Consensus Project, the goal of palliative care is to prevent and relieve suffering and to support the best possible quality of life for patients and their families, regardless of the stage of the disease or the need for other therapies.

Palliative care expands traditional disease-modifying medical treatments to include the goals of:

- Enhancing quality of life for patient and family
- Optimizing function
- Assisting with decision making
- Providing opportunities for personal growth.

Palliative care can be delivered concurrently with life-prolonging care or as the main focus of care.

A Comprehensive Model of Palliative Care

Risk

Disease-modifying therapy
(Curative, Restorative)

Life Closure

Palliative Care

Disease

Condition

Death & Bereavement

National Hospice Work Group. Adapted from work of the Canadian Palliative Care Association & Frank Ferris, MD

#ChooseKnowledge
The American Academy of Pediatrics Policy on Palliative Care for Children advocates for an integrated model of palliative care “in which the components of palliative care are offered at diagnosis and continued throughout the course of illness.”


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Palliative Care

- Therefore, palliative care is a comprehensive approach to care that focuses on the treatment of physical, emotional, social, and spiritual symptoms of children with life-threatening conditions and their families.
- This care can, and should be provided concurrently with curative or life-prolonging care.
- The goal of palliative care is to achieve the best quality of life for children and their families.

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Early palliative care

- Patients with newly diagnosed metastatic non–small-cell lung cancer were randomly assigned to receive either early palliative care integrated with standard oncologic care or standard oncologic care alone.
- Quality of life and mood were assessed at baseline and at 12 weeks with the use of the Functional Assessment of Cancer Therapy–Lung (FACT-L) scale and the Hospital Anxiety and Depression Scale.
- The primary outcome was the change in the quality of life at 12 weeks.


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Early palliative care

- 151 patients were enrolled and underwent randomization
- 74 received standard care alone / 77 received standard care plus early palliative care
- 27 died by 12 weeks
- 107 (86%) completed assessments.
- There were no differences in baseline characteristics between the two groups.
- A trend toward a higher proportion of minority patients in the standard care arm

#ChooseKnowledge

Early palliative care – Quality of Life

<table>
<thead>
<tr>
<th>Variable</th>
<th>Standard Care (N=47)</th>
<th>Early Palliative Care (N=66)</th>
<th>Difference between Early Care and Standard Care (95% CI)</th>
<th>P Value</th>
<th>Effect Size</th>
</tr>
</thead>
<tbody>
<tr>
<td>FACT-G</td>
<td>93.5±15.8</td>
<td>98.6±15.3</td>
<td>5.1 (0.5-10.4)</td>
<td>0.09</td>
<td>0.42</td>
</tr>
<tr>
<td>LCS score</td>
<td>15.3±4.2</td>
<td>21.6±3.9</td>
<td>6.3 (1.5-10.4)</td>
<td>0.009</td>
<td>0.52</td>
</tr>
<tr>
<td>TDA score</td>
<td>59.6±11.5</td>
<td>59.6±11.4</td>
<td>0.0 (1.5-1.6)</td>
<td>0.41</td>
<td>0.41</td>
</tr>
</tbody>
</table>

#ChooseKnowledge

Early palliative care – Depression

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Early palliative care – Survival

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#ChooseKnowledge
Goals of care

- For as long as the practice of medicine has existed, there have been two primary goals of care: 1) to cure disease and 2) to relieve suffering in patients.
- Within those two broad categories, there are many specific goals unique to each individual patient that will help satisfy the care mission.
- Palliative care seeks to relieve the physical, emotional, social, and spiritual distress produced by life-limiting conditions, to assist in complex decision-making, and to enhance the quality of life.

Emanuel LL. Goals of care. In: The Education for Physicians on End-of-life Care (EPEC) curriculum, Module 7; Institute for Ethics at the American Medical Association; 1999.

Goals of care

- Providers must guide realistic goal setting for the patient.
- Many publications, as well as national guidelines, have reaffirmed the importance of developing goals of care.
- The Medicare Hospice Benefit has long contained a provision mandating individualized treatment plans including the establishment of goals of care for patients.
- These goals of care should be rooted in the personal beliefs of the patient and family and based on the clinical condition.
- The goals of the child should be solicited and respected.

Goals of care

- Data suggest that most patient goals can be categorized into general areas pertaining to quality of life.
- World Health Organization. WHOQOL-BREF
- This categorization may assist families in establishing more broad-based goals of care.
- Goals of care often remain poorly delineated because they are not discussed thoroughly and openly.
- Pediatric nurses rated "uncertainty about the goals of care" as second only to "lack of opportunity to debrief after death", in a study of obstacles to the provision of palliative care.
Goals of care

- Goals of care were elicited from the parents and children with complex, life-limiting conditions during initial palliative care consultation.
- Data abstracted included: diagnoses, demographics, time from diagnosis until initial palliative care consult, spirituality status, resuscitative status, and disposition at discharge.
- Goals of care were categorized into one of four quality-of-life domains:
  - Physical health and independence
  - Social
  - Psychological and spiritual
  - Environment

One hundred and forty goals of care were obtained from 50 patients/parents.
- The median patient age was 4.6 years.
- Forty-nine patients identified at least one goal pertaining to physical health and independence.
  - Significantly more than any other category (p<0.0001)
  - 25 verbalized a goal that specifically and directly addressed health maintenance or improvement
Goals of care

Survey of 103 parents of children who died of cancer

- During the parent-defined end-of-life care period
  - Two thirds of parents voiced that the primary goal of cancer-directed therapy was to extend life
  - Specifically to cure in over a quarter of the cases


Goals of care

- Pediatric patients 10 or more years of age and a parent were interviewed within 7 days of participating in one of the following three end-of-life decisions:
  - Enrollment onto a phase I trial (n = 7)
  - Adoption of a do not resuscitate order (n = 5)
  - Initiation of terminal care (n = 8)
- Twenty patients participated
  - Ages 10 to 20 years (mean, 17 years and 4 months)
  - Diagnoses included refractory solid tumor (n = 12), brain tumor (n = 4), or leukemia (n = 4) participated

Goals of Care

As such, one of the primary tasks of the palliative care clinician is to meet with the patient and family and establish realistic goals of care. As goals will vary from patient to patient, it is not the role of the clinician to determine what these goals should be. That is the role of the patient and/or the family. However, the palliative care team must meet with them to assist in the development and implementation of their goals.

Interesting to note, in a recent report of 50 children referred for palliative care consultation, nearly all (98%) patient/family units expressed at least one goal pertaining directly to physical health and independence.

This finding is important for two reasons: First, it re-asserts that patients/families concur with the philosophy that palliative care should be administered concurrently with traditional medical care. Second, it emphasizes the need to not eliminate hope as families clearly have hope for physical health gains or maintenance. Instead, it is important to channel that hope to realistic goals.

Goals of care

- Within the realm of physical health, common goals expressed by palliative care patients other than restoration of health often include those pertaining to symptom management and maintenance of independence.
- 24 of 50 patients/families during initial palliative consult verbalized the specific goals "To be comfortable" and/or "To not suffer."
- An additional six of these patients/families verbalized a goal "To be independent."
- Treatment of symptoms and interventions to foster and maintain independence are well established cornerstones of palliative care.

Prevention and identification of suffering


"Fundamentals of Pediatric Palliative Care"
Robert F. Tamburro, MD
**Physiology**

**Brain**
- Brainstem medial & lateral reticular formations
- Medullary raphe nuclei
- Periaqueductal gray
- Thalamus
- Cerebral cortex

**Peripheral Pain Receptor Responsiveness**
- PG, serotonin, H+, Bradykinin, NE, K+, Substance P

**Substance P** facilitates nociceptive transmission at the interneurons

**Opioid Receptors**
- Innocuous, non-painful stimuli

**Dorsal Root Ganglia**
- Spinothalamic spinoreticular pathways

**Anxiety, fear, age, helplessness, affect, culture, sleeplessness, previous pain, socioeconomic status**

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**Pain management in palliative care**
- Multi-dimensional approach required
- Pharmacologic therapy
  - Nonsteroidal anti-inflammatory agents
  - Opioids
  - Adjuvants
- Non-pharmacologic therapy

Effective pain management usually requires a multi-dimensional approach utilizing both pharmacologic and non-pharmacologic interventions.

**Nonsteroidal medications**
- Nonsteroidal medications are used for mild pain and as an analgesic adjuvant.
- They are particularly useful in the setting of bone pain.
- Common examples include acetaminophen, naproxen, ibuprofen, trilisate, and celecoxib
- All should be used with caution in the setting of renal dysfunction, with concomitant diuretic use or with hypovolemia.
- They are characterized by ceiling effects meaning that above a certain dose all that is obtained is toxicity without additional beneficial effect.
Opioids may be used to treat mild to moderate pain. A weak opioid may be used in conjunction with a nonsteroidal agent.

Examples of weak opioids include each of the following:
- Hydrocodone
- Oxycodone
- Codeine
- Propoxyphene

Meperidine should be avoided because of its poor safety profile, and the availability of other effective agents.

More potent opioids are utilized to effectively treat moderate to severe pain.

Examples of such opioids include each of the following:
- Morphine
- Hydromorphone
- Methadone
- Fentanyl
- Oxycodone

The oral route is always the preferred route of opioid delivery.
- This mode of delivery is effective, convenient, and less expensive than other modes.
- It may also be associated with fewer serious side effects.
- If the oral route is not tolerated, there are many other modes of opioid delivery:
  - Intravenous
  - Transcutaneous
  - Subcutaneous
  - Transmucosal (buccal)
Dyspnea

In addition to pain prevention and treatment, adequate control of dyspnea is paramount to effective end-of-life care.

Dyspnea has been defined as the subjective awareness of uncomfortable breathing.

It is usually characterized as a symptom and nociceptive phenomenon in response to an aversive stimulus.

It may be very distressing to witness.

It requires intensive management and reassessment.

It is usually attributed to the underlying disease process.

It may be associated with multiple etiologies which can be treated.

Causes of Dyspnea

<table>
<thead>
<tr>
<th>Causes</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bronchospasm</td>
<td>B-agonists; Steroids</td>
</tr>
<tr>
<td>Rales</td>
<td>Fluid restriction; Diuretics; Antibiotics</td>
</tr>
<tr>
<td>Effusions</td>
<td>Thoracentesis; Pleurodesis</td>
</tr>
<tr>
<td>Airway Obstruction</td>
<td>Thickenfeeds (Aspiration)</td>
</tr>
<tr>
<td>Thick Secretions</td>
<td>+ Cough Reflex: Nebulized Saline</td>
</tr>
<tr>
<td></td>
<td>- Cough Reflex: Anticholinergics</td>
</tr>
<tr>
<td>Low Hemoglobin</td>
<td>Blood</td>
</tr>
<tr>
<td>Anxiety</td>
<td>Relaxation; Opioids / Benzodiazepines</td>
</tr>
<tr>
<td>Interpersonal Concerns</td>
<td>Multidisciplinary Team</td>
</tr>
<tr>
<td>Religious Concerns</td>
<td>Spiritual Services</td>
</tr>
</tbody>
</table>

General Treatment of Dyspnea

If the benefit of treatment is greater than the burden of treatment

Palliative therapy should be used if:

No treatable cause is identified

The burden of investigation or treatment of dyspnea is greater than the benefit of treatment

If treatment does not completely alleviate the symptoms

Treat the underlying cause

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Management of Dyspnea

- Non pharmacologic therapies
- Pharmacologic therapies

Non Pharmacologic Management of Dyspnea

- Effective positioning
- Non-invasive ventilation (BiPAP, CPAP)
- Relieve anxiety
- Avoid strong odors

Effective positioning

Fans are a great way to improve air circulation. In fact, the air stimulates the V2 branch of the 5th cranial nerve which centrally inhibits the sensation of breathlessness.

Non-invasive ventilation (BiPAP, CPAP)

In addition to pharmacologic therapy, anxiety may be reduced by providing reassurance. Relaxation techniques may also be very helpful.

Relieve anxiety

Avoid strong odors

It is important to realize that fatigue, nausea and emesis are other symptoms that impair quality of life. In brief, in treating nausea/emesis, it is important to determine the etiology of the nausea. Different etiologies will mediate the symptoms through different pathophysiological mechanisms, and to some degree through different neurotransmitters. Consequently, different medications may be more beneficial in specific symptoms.
As an example, post-operative nausea and vomiting is mediated by the release of serotonin from enterochromaffin cells in the stomach and intestine depicted in the photograph insert.

Post-operative nausea and vomiting

Enterochromaffin cells releasing serotonin

Vagal Nerve

Serosa

GI tract

Stomach and stomach lining

Enterochromaffin cells releasing serotonin

Serotonin

5-hydroxytryptamine type 3 (5-HT3) receptors

Chemoreceptor Trigger Zone

VOMIT

Vagal nerve

Serotonin binds to (5-HT3) receptors in the GI tract. This binding results in stimulation of vagal afferents in the GI tract that conduct impulses reaching brainstem structures located on the dorsal surface of the medulla at the caudal end of the fourth ventricle, the area postrema. This area of the brain, the chemoreceptor trigger zone, has a critical role in the central mechanism of vomiting. As such, it is easy to understand why 5-HT3 receptor antagonists such as ondansetron are useful in post-operative nausea and vomiting.

In contrast, nausea and vomiting associated with perturbations of the vestibular and limbic system such as motion sickness are mediated by acetylcholine. Therefore, anticholinergic medications such as scopolamine are much more likely to be effective for nausea and emesis in this setting.
Bioethics of Palliative Care

- Do Not Attempt Resuscitation
- Differences between Withdrawal and Limiting of Life-sustaining Therapies
- The Doctrine of Double Effect
- Conflict over Futile or Potentially Inappropriate Therapies
- Brain Death and Organ Donation