Fundamentals of Pediatric Palliative Care

Pennsylvania Osteopathic Medical Association
111th Annual Clinical Assembly and Scientific Seminar

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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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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.

Cunningham RM. *N Eng J Med* 2018; 379:2468-2475.
Hoyert DL. *Pediatrics* 2006; 117:168.

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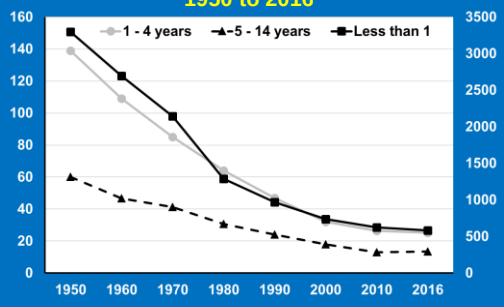
Mortality rates for three US pediatric cohorts 1950 to 2016



Suttle ML. *Pediatr Clin North Am.* 2017;64(5):1167-1183.

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Epidemiology

TABLE 1 Site of Death Varies by Age

Age	Site of Death Frequency, %				Total
	Hospital	Nursing Facility	Home	Emergency Department	
<1 mo	3527 (92.9)	7 (0.2)	185 (4.9)	77 (2.0)	3796 (100)
1–11 mo	1145 (45.6)	8 (0.3)	864 (34.4)	493 (19.6)	2510 (100)
1–9 y	1060 (53.4)	15 (0.8)	632 (31.8)	279 (14.1)	1986 (100)
10–19 y	1473 (53.3)	23 (0.8)	864 (31.3)	405 (14.7)	2765 (100)
20–39 y	7770 (44.9)	1135 (6.6)	7062 (40.8)	1348 (7.8)	17 315 (100)
40–59 y	27 141 (45.2)	5624 (9.4)	24 459 (40.7)	2846 (4.7)	60 070 (100)
60–79 y	87 539 (42.7)	44 640 (21.8)	66 958 (32.7)	5803 (2.8)	204 940 (100)
>79 y	60 227 (27.6)	108 872 (49.9)	46 379 (21.2)	2931 (1.3)	218 409 (100)
Total	189 882 (37.1)	160 324 (31.3)	147 403 (28.8)	14 182 (2.8)	511 791 (100)

Feudtner C. *Pediatrics* 2006; 117:e932.

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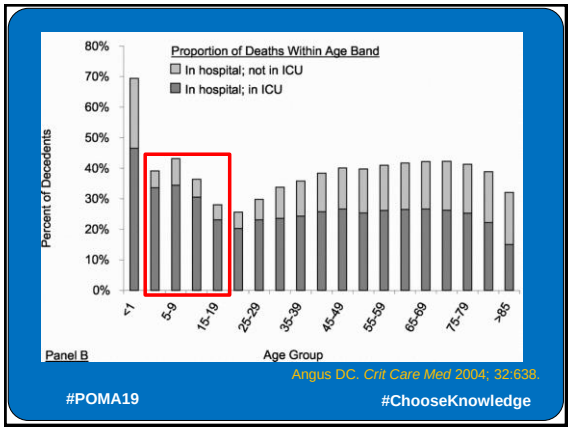

Angus DC. *Crit Care Med* 2004; 32:638.

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TABLE 1. Sociodemographics of Children Whose Families Did and Did Not Discuss Limitation or Withdrawal of Life Support With Clinicians

Sociodemographics	Did Not Have Discussions n = 9,830 n (%)	Had Discussions n = 348 n (%)	p*
Gender			0.193
Male	5,404 (55.0)	126 (50.8)	
Female	4,426 (45.0)	122 (49.2)	
Age at PICU admission			< 0.001
0 d to < 14 d	455 (4.6)	37 (14.9)	
14 d to < 1 mo	137 (1.4)	1 (0.4)	
1 mo to < 12 mo	2,111 (21.5)	53 (21.4)	
> 12 mo	7,127 (72.5)	157 (63.3)	
Race/ethnicity			< 0.001
Hispanic	1,656 (16.8)	70 (28.2)	
American Indian or Alaska Native, NH	135 (1.4)	7 (2.8)	
Asian, NH	253 (2.6)	10 (4.0)	
Black or African American, NH	2,231 (22.7)	40 (16.1)	
Native Hawaiian or other Pacific Islander, NH	34 (0.3)	2 (0.8)	
White, NH	4,622 (47.0)	89 (35.9)	
Multiracial, NH	14 (0.1)	0 (0)	
Unknown or not reported	885 (9.0)	30 (12.1)	
Payer type			< 0.001
Government	5,248 (53.4)	172 (69.4)	
Nongovernment	4,358 (44.3)	70 (28.2)	
Missing	224 (2.3)	6 (2.4)	

Keele L. Pediatr Crit Care Med. 2016;17:110-120.

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TABLE 2. Clinical Characteristics at PICU Admission for Children Whose Families Did and Did Not Discuss Limitation or Withdrawal of Life Support With Clinicians

Clinical Characteristics	Did Not Have Discussions n = 9,830 n (%)	Had Discussions n = 348 n (%)	p*
Primary diagnosis			< 0.001
Respiratory	3,285 (33.4)	91 (26.7)	
Cancer	359 (3.7)	11 (4.4)	
Cardiovascular (acquired)	624 (6.3)	49 (19.8)	
Cardiovascular (congenital)	1,706 (17.4)	51 (20.6)	
Neurologic	1,987 (20.2)	35 (14.1)	
Miscellaneous†	1,869 (19.0)	11 (4.4)	
Primary or secondary diagnosis of trauma	635 (6.5)	15 (6.0)	0.795
Chronic diagnoses at admission	7,213 (73.4)	201 (81.0)	0.01
Known developmental delay	2,373 (24.1)	98 (39.5)	< 0.001
Baseline Functional Status Scale			< 0.001
Good (6, 7)	7,099 (72.2)	134 (54.0)	
Mildly abnormal (8, 9)	996 (10.1)	28 (11.3)	
Moderately abnormal (10–15)	1,216 (12.4)	53 (21.4)	
Severely abnormal (16–21)	397 (4.0)	20 (8.1)	
Very severely abnormal (> 21)	122 (1.2)	13 (5.2)	
Baseline Pediatric Overall Performance Category			< 0.001
Good	3,847 (39.1)	69 (27.8)	
Mild disability	3,170 (32.2)	41 (16.5)	
Moderate disability	1,818 (18.5)	59 (23.8)	
Severe disability	899 (9.1)	71 (28.6)	
Coma/vegetative	96 (1.0)	8 (3.2)	
Baseline Pediatric Cerebral Performance Category			< 0.001
Normal	6,827 (69.5)	132 (53.2)	
Mild disability	1,547 (15.7)	33 (13.3)	
Moderate disability	732 (7.4)	27 (10.9)	
Severe disability	631 (6.4)	48 (19.4)	
Coma/vegetative	93 (0.9)	8 (3.2)	
Worst Glasgow Coma Scale			< 0.001
8–15	9,261 (94.2)	163 (65.7)	
3–7	565 (5.7)	85 (34.3)	
Unable to assess	4 (0)	0 (0)	

#POMA19 Keele L. Pediatr Crit Care Med. 2016;17:110-120.

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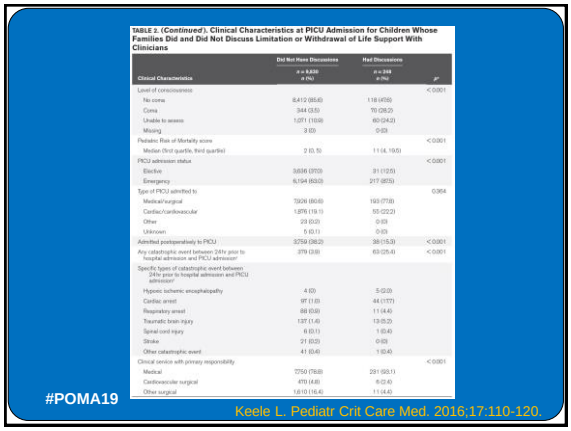
TABLE 2. (Continued). Clinical Characteristics at PICU Admission for Children Whose Families Did and Did Not Discuss Limitation or Withdrawal of Life Support With Clinicians
#POMA19
Keele L. Pediatr Crit Care Med. 2016;17:110-120.

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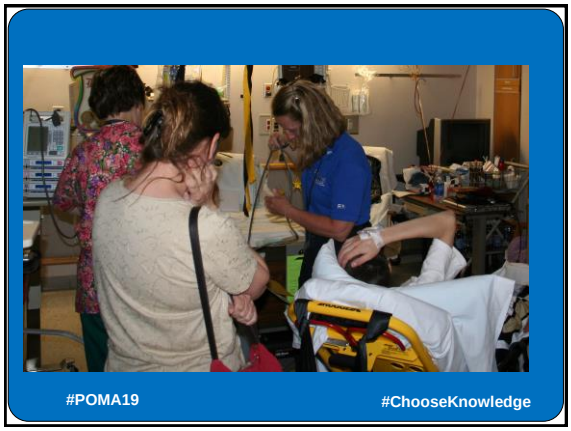
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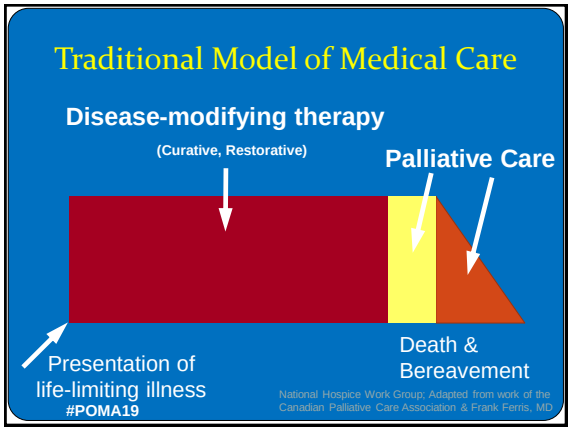
Traditional Model of Medical Care
Disease-modifying therapy
(Curative, Restorative)
Palliative Care
Presentation of life-limiting illness
Death & Bereavement
National Hospice Work Group; Adapted from work of the Canadian Palliative Care Association & Frank Ferris, MD
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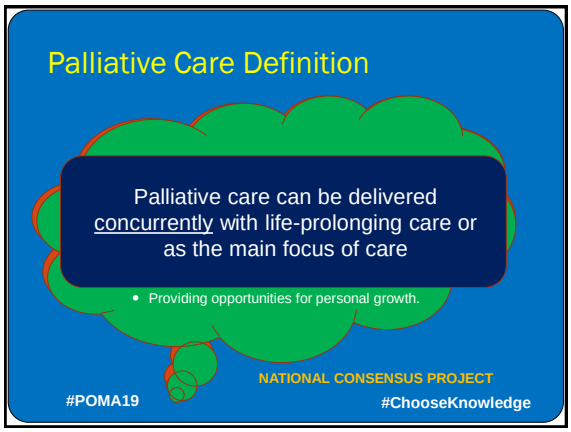
Palliative Care Definition
Palliative care can be delivered concurrently with life-prolonging care or as the main focus of care
• Providing opportunities for personal growth.
NATIONAL CONSENSUS PROJECT
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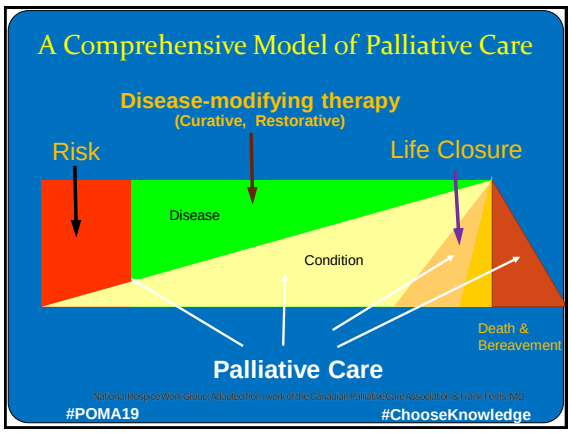
A Comprehensive Model of Palliative Care
Disease-modifying therapy
(Curative, Restorative)
Risk
Life Closure
Disease
Condition
Death & Bereavement
Palliative Care
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The American Academy of Pediatrics Policy on Palliative Care for Children advocates for an integrated model of palliative care "*in which the components of palliative care are offered at diagnosis and continued throughout the course of illness*."

Field MJ. When children die: improving palliative and end-of-life care for children and their families. Institute of Medicine. Washington, DC: The National Academies Press; 2003.

American Academy of Pediatrics: Committee on Bioethics and Committee on Hospital Care: Palliative care for children. Pediatrics 2000;106:351–357.

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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.

Temel JS. *N Eng J Med* 2010; 363:733-42.

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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

Temel JS. *N Eng J Med* 2010; 363:733-42.

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Early palliative care – Quality of Life

Table 2. Bivariate Analyses of Quality-of-Life Outcomes at 12 Weeks.*

Variable	Standard Care (N = 47)	Early Palliative Care (N = 60)	Difference between Early Care and Standard Care (95% CI)	P Value†	Effect Size‡
FACT-L score	91.5±15.8	98.0±15.1	6.5 (0.5–12.4)	0.03	0.42
LCS score	19.3±4.2	21.0±3.9	1.7 (0.1–3.2)	0.04	0.41
TOI score	53.0±11.5	59.0±11.6	6.0 (1.5–10.4)	0.009	0.52

Temel JS. *N Eng J Med* 2010; 363:733-42.

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Early palliative care – Depression

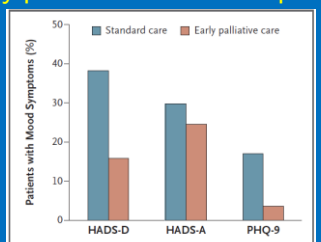

Figure 2. Twelve-Week Outcomes of Assessments of Mood.

Temel JS. *N Eng J Med* 2010; 363:733-42.

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Early palliative care – Survival

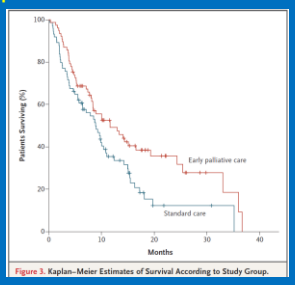

Figure 3. Kaplan–Meier Estimates of Survival According to Study Group.

Temel JS. *N Eng J Med* 2010; 363:733-42.

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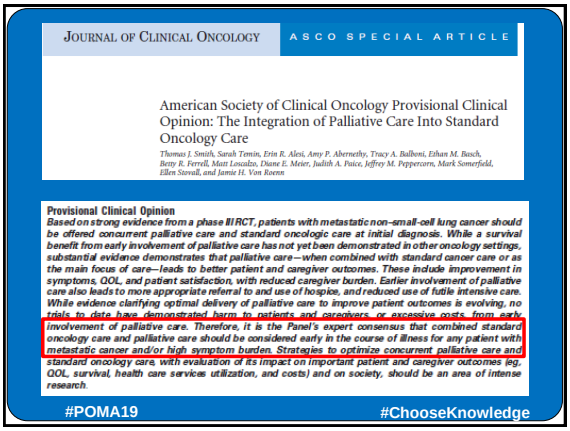

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Goals of care

- 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.

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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.

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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.
  - Feudtner C. Pediatr Clin North Am 2007;54:583–607.
- 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.
  - Tibbs-Cooley HL. J Palliat Med 2011;14:631-37.

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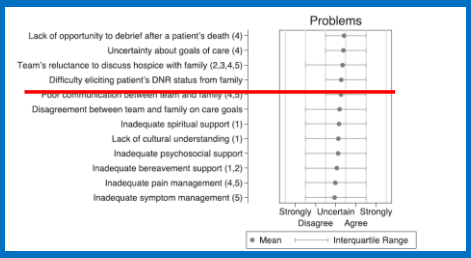
Goals of care



Tubbs-Cooley HL. J Palliat Med 2011;14:631-37

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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

Tamburro RF. J Palliat Med 2011;14:607-613

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Goals of care

- 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

Tamburro RF. J Palliat Med 2011;14:607-613

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Goals of care

- Survey of 103 parents of children who died of cancer
- During the *parent-defined* end-of-life care period
  - Ttwo 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

Wolfe J. JAMA 2000;284:2469-75.

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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

Hinds PS. J Clin Oncol 2005;23:9146-54.

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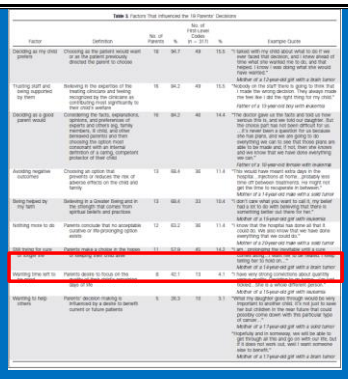

Hinds PS. J Clin Oncol 2005;23:9146-54.

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Goals of Care

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.

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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.

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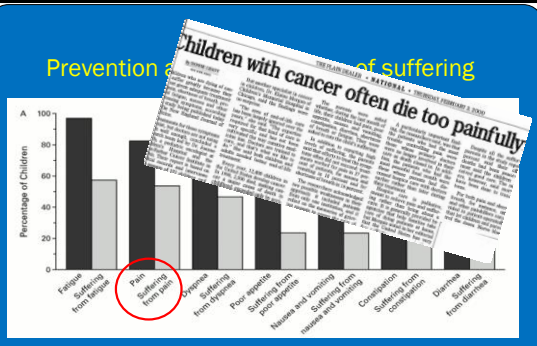
Prevention a of suffering



Wolfe J. *NEJM* 2000; 342:326.

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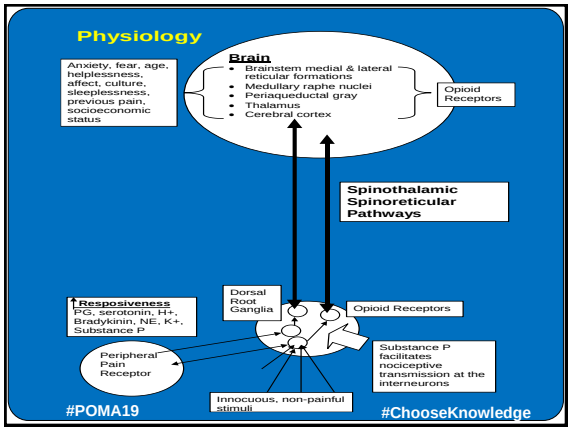
Physiology
Anxiety, fear, age, helplessness, affect, culture, sleeplessness, previous pain, socioeconomic status
Brain
Brainstem medial & lateral reticular formations
Medullary raphe nuclei
Periaqueductal gray
Thalamus
Cerebral cortex
Opioid Receptors
Spinothalamic Spinoreticular Pathways
Responsiveness PG, serotonin, H+, Bradykinin, NE, K+, Substance P
Dorsal Root Ganglia
Opioid Receptors
Peripheral Pain Receptor
Substance P facilitates nociceptive transmission at the interneurons
Innocuous, non-painful stimuli
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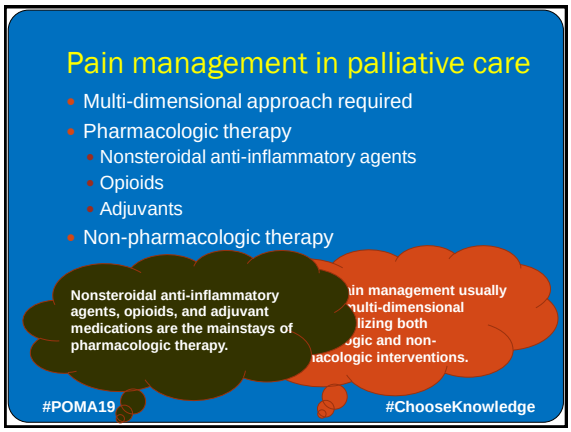
Pain management in palliative care
Multi-dimensional approach required
Pharmacologic therapy
Nonsteroidal anti-inflammatory agents
Opioids
Adjuvants
Non-pharmacologic therapy
Nonsteroidal anti-inflammatory agents, opioids, and adjuvant medications are the mainstays of pharmacologic therapy.
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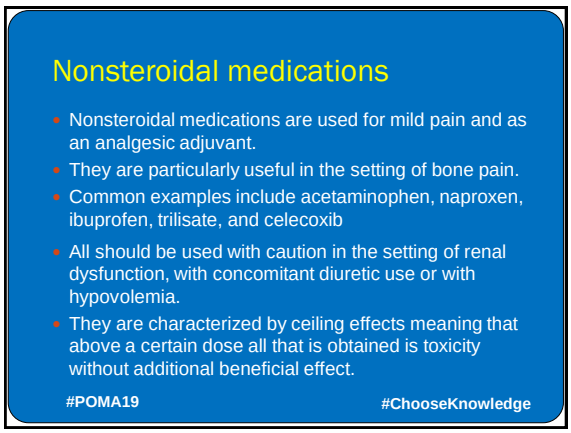
Nonsteroidal medications
Nonsteroidal medications are used for mild pain and as an analgesic adjuvant.
They are particularly useful in the setting of bone pain.
Common examples include acetaminophen, naproxen, ibuprofen, trilisate, and celecoxib
All should be used with caution in the setting of renal 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.
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Opioids

- 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.

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Opioids

- 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

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Opioids – General dosing principles

- 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)

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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.

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	Causes of Dyspnea	Treatment
B	Bronchospasm	*B*-agonists; Steroids
R	Rales	Fluid restriction; Diuretics; Antibiotics
E	Effusions	Thoracentesis; Pleurodesis
A	Airway Obstruction	Thicken Feeds (Aspiration)
T	Thick Secretions	+ Cough Reflex: Nebulized Saline - Cough Reflex: Anticholinergics
H	Low Hemoglobin	Blood
A	Anxiety	Relaxation; Opioids / Benzodiazepines
I	Interpersonal Concerns	Multidisciplinary Team
R	Religious Concerns	Spiritual Services

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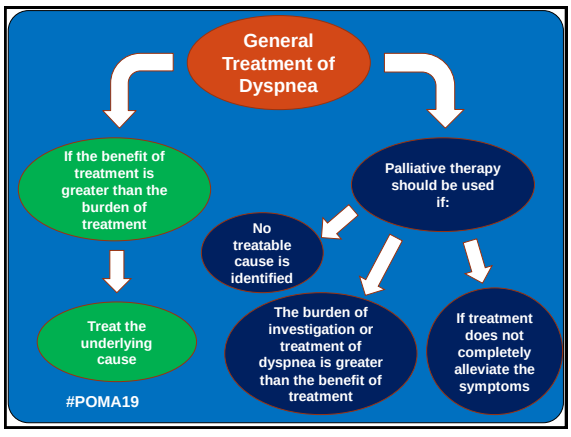

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Management of Dyspnea

- Non pharmacologic therapies
- Pharmacologic therapies

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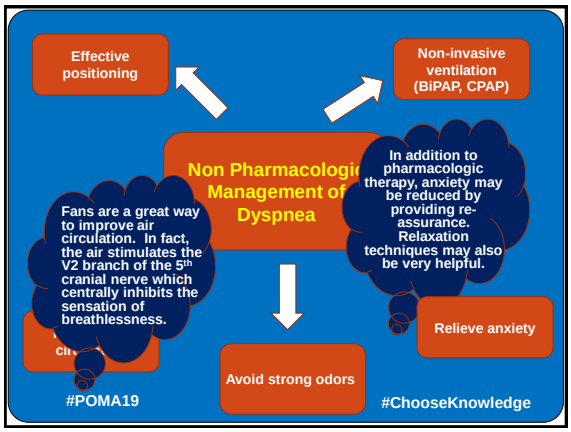

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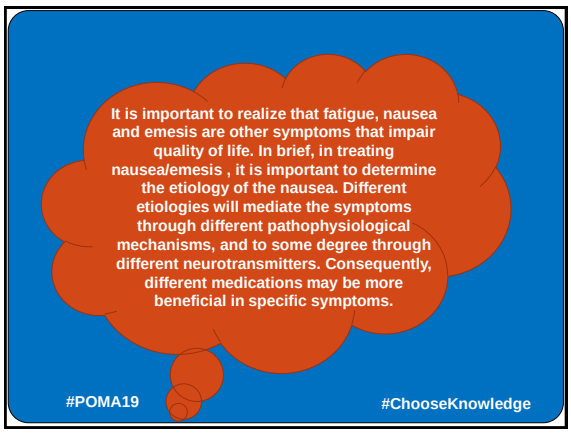
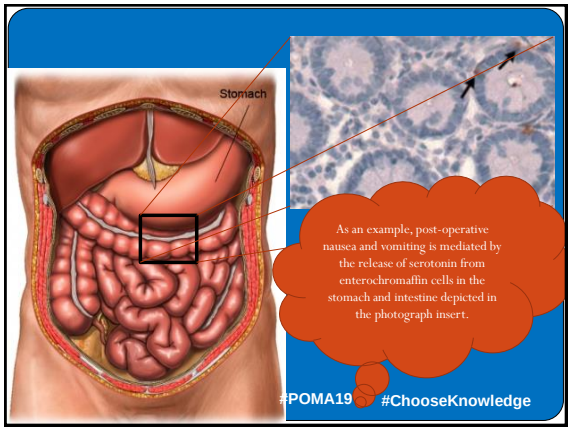


48


Stomach
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.
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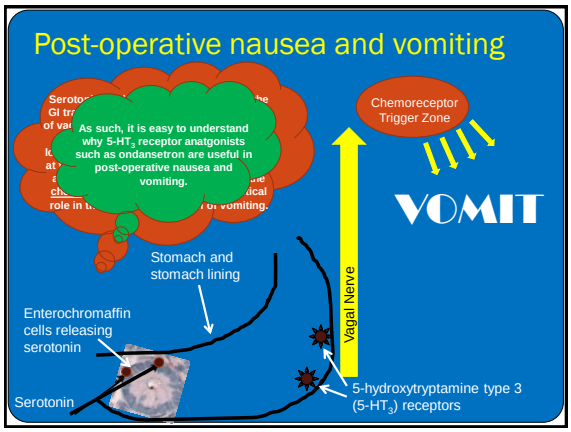
Post-operative nausea and vomiting
As such, it is easy to understand why 5-HT3 receptor anatgonists such as ondansetron are useful in post-operative nausea and vomiting.
Chemoreceptor Trigger Zone
VOMIT
Stomach and stomach lining
Enterochromaffin cells releasing serotonin
Serotonin
Vagal Nerve
5-hydroxytryptamine type 3 (5-HT3) receptors

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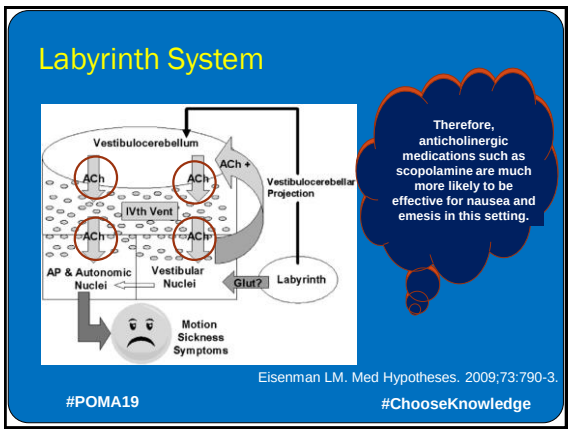
Labyrinth System
Vestibulocerebellum
ACh
ACh +
Vestibulocerebellar Projection
IVth Vent
AP & Autonomic Nuclei
Vestibular Nuclei
Glut?
Labyrinth
Motion Sickness Symptoms
Therefore, anticholinergic medications such as scopolamine are much more likely to be effective for nausea and emesis in this setting.
Eisenman LM. Med Hypotheses. 2009;73:790-3.
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General approach to nausea and vomiting in the pediatric palliative care patient.

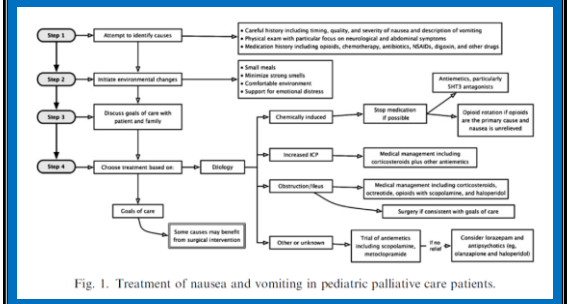

Fig. 1. Treatment of nausea and vomiting in pediatric palliative care patients.

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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

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