



NCC Pediatrics Continuity Clinic Curriculum: Palliative & End-of-Life Care *Faculty Guide*

Goals & Objectives:

To evaluate and discuss pain and other symptoms at the end of life:

- Identify common symptoms associated with the final stages of dying and describe the importance of immediate treatment.
- Recognize the importance of empowering children and parents to make choices regarding day-to-day as well as long-term concerns regarding the child's treatment.
- Identify the types of choices and range of options that are available or could be implemented in your hospital to increase the voice of children and families in the organization and implementation of family-centered pediatric palliative care.

Pre-Meeting Preparation:

Please choose 1-2 of the following enclosures/links to review:

- **Video Links:** [American Cancer Society](#); [NIH/NINR](#) (2-4 min each)
 [“A Little Warrior’s Journey”](#) (video by Dr. Eberly)
- Pediatric Palliative Care Checklist
- “Pediatric Palliative, End-of-Life, and Bereavement Care” (2005)
 - *Review Table 5: Self-assessment questions*

Conference Agenda:

- Complete [“My Wishes”](#) workbook (*hard copies will be provided*). **Reflect as a group upon this exercise, as well as the video links your group watched.**
 - Optional [Discussion Questions](#) for “A Little Warrior’s Journey” video
- Review Palliative Care Quiz

Extra-Credit:

- [IPPC: Initiative for Pediatric Palliative Care](#): *Online curriculum for providers.*
- [AAP Section on Hospice & Palliative Care](#): *Multiple links for providers & parents*
- **Palliative & EOL Care Research:**
 - [“Pediatric Palliative Care”](#) (*PIR, 2007*)—*review article*
 - [“Pediatric Palliative Care”](#) (*NEJM, 2004*)—*review article*
 - [“Collaborative Communication”](#) (*Ped Clin NA, 2007*)—*framework for discussion*
- **Parent Resources:**
 - [“A Lion in the House”](#): *Documentary + parent guide. **15 helpful links at end.*
 - [Bereavement Services & Funeral Homes in National Capital Area](#)

Palliative Care Plan & Checklist

(Developed by Dr. Jason Higginson)

Goal Setting Discussion Points:

1. Understanding of progression of disease? (*allow them to state in own words*)
2. What goals does child/family have for the child's life and of treatment?
3. Discuss spirituality/religious beliefs (*how these relate to beliefs about health care and death and dying*).
4. Does the family know how to handle emergencies? What possible emergencies can arise and what do they want done?
5. When to call the doctor or hospital.
6. What to expect as child approaches end of life: *signs and symptoms of impending death*.
7. Where do they desire death to occur: *home, hospital, hospice or other location*?
8. What to expect when the child dies (i.e. *physical, emotional*). What dead body looks like, sounds, smells, etc.
9. Have they started to consider funerals/funeral home-arrangements? (*Make prior to death if possible*). If death is to be at home, arrangements for pick-up, what to do if child dies at home (*who needs to declare child, etc.*).
10. Does family have written copy of what an advanced directive? DNR/DNI?
11. What to do if child becomes distressed or unconscious at home?

Identification of resources: *What resources do they currently have or know about?*

- Palliative care services
- Physician
- Nurse
- Volunteers
- Respite

Preferences for on-going care:

- Blood work
- PE/Check-up interval desired
- Clinic/office visits
- IV fluid support
- Nutrition support
- Breathing support
- Infections/antibiotics
- Blood/product transfusions
- DNR
- Pain medication

Expected symptoms & their management:

- Respiratory
- Pain
- Constipation
- Diarrhea
- Anorexia
- N & V
- Bleeding:
- Anemia
- Infection
- Seizures
- Neurological

Pediatric Palliative, End-of-Life, and Bereavement Care

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Each year in the United States, approximately 50,000 children die, mostly in hospitals, and many more confront life-threatening conditions. Most—if not all—of these children and their families would benefit from timely, comprehensive, compassionate, continuous, and developmentally appropriate supportive care services, including a mixture of palliative, end-of-life, and bereavement care. [1,2]

Palliative care aims to relieve suffering and improve quality of life for patients with advanced illnesses and their families. Effective palliative care includes communicating with patients and family members, managing pain and other symptoms, providing psychosocial, spiritual, and bereavement support, and

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coordinating various medical and social services. For dying people of all ages, care should be directed toward relieving symptoms and suffering. For children, such care can be instituted in conjunction with curative therapy. Palliative care is appropriate for children with a wide range of conditions, even when cure remains a distinct possibility. The American Academy of Pediatrics supports the concepts of palliative care, stating that “the components of palliative care are offered at diagnosis and continued throughout the course of illness, whether the outcome ends in cure or death. Palliative care should be accessible in any setting, including home, hospital, and school” [3]. End-of-life care is more narrowly conceived as a phase of palliative care that is provided to patients who are manifesting the signs and symptoms of dying or who require certain procedures to be performed, such as the medical task of ceasing mechanical ventilation or the religious ceremony of last rites. Bereavement care addresses the experience and consequences of grief, which can be manifest before a child dies (so-called anticipatory grief) and after a child’s death. Although these three modes of care can be discussed as if they were distinct, in practice they are inexorably entwined.

Historically, children with advanced illness and their families have not received adequate palliative, end-of-life, and bereavement care, partly because of a medical philosophy focused on cure rather than quality of life and a tendency to focus on quality of life only when death is imminent. We realize, however, that children benefit most from care that includes a combination of life-prolonging treatment, palliation of symptoms, rehabilitation, and support for caregivers.

The pediatric hospitalist plays an integral role in providing palliative, end-of-life, and bereavement care for children and families. This article focuses on a multifaceted approach to palliative, end-of-life, and bereavement care, in which the physician is a key member of an interdisciplinary team. We believe that we can improve quality of life and relieve suffering only by paying attention to the medical, emotional, spiritual, and practical needs and goals of dying children and their loved ones [4,5].

Talking to families

Communication, the great facilitator of human relationships, is the foundation of palliative care [6]. Good communication enables patients and family members to collaborate effectively with health care providers and allows these providers to support patients and family as they make decisions and confront the reality of their circumstances. Conversely, bad communication impairs the quality of care and can create negative memories that linger in the minds of family members for years.

Parents of sick children value health care providers who communicate clearly, accurately, and empathically. Importantly, these characteristics are based in part on attributes that individual health care providers possess. Not only is a solid font of knowledge essential but also the provider must be mentally and emotionally

prepared for and focused on the task. Equally essential is the ability to devise personal ways to “step out of” the hectic pace of hospital practice and enter a quieter and more intimate mode of relating to other people.

Clarity, accuracy, and empathy in communication also result from specific techniques or habits of how to have difficult discussions. Table 1 outlines steps that are useful when the principal task involves breaking bad news [7]. One must realize that “bad” news can range from the diagnosis of a fatal condition to the message that discharge to home will be delayed because of difficulty staffing home nursing. Once a family has assimilated the news and its implications,

Table 1
Breaking bad news

Steps	Comment
Make a plan	Providers too often initiate a conversation or family meeting without a clear concept of what they hope to achieve; before the discussion, take a minute to mentally rehearse the steps outlined later
Getting the setting and people right	A quiet, private room is best; make sure that all the necessary people — as defined by the family — are present and that you allot enough time
Briefly recap the situation	Confirm that all the people assembled have the same understanding of the situation; state the fundamental issues confronting the child and any pending test results or other events that have prompted the meeting
Provide a warning shot	Before delivering the bad news, allow the parents a few seconds to prepare themselves by uttering a short phrase, such as “I wish the results were different, but . . .” or “I am afraid that what we were worried about has happened, and . . .”
State the bad news simply	Deliver the bad news in plain, straightforward language: “. . . but the test shows that the cancer has come back” or “and our effort to use the machine to breathe for your child is no longer working”
Allow silence	After the news is delivered, silence often fills the room; avoid the temptation to fill the silence immediately with talk; be quiet for a minute
Acknowledge emotion	A simple, humble statement, such as “I can’t imagine what you are feeling,” can be helpful in addressing one of the most palpable aspects of this experience
Answer questions	Minds often go blank after receiving bad news; solicit questions after recognizing this reality: “I realize that this news is overwhelming. I’m dedicated to helping you understand what it means to your child. If you have questions now or later, once you’ve thought about this for a while, I’m here and will be here to answer those questions.”
Formulate a next-step plan	Depending on how unexpected the news is, now may or may not be the time to discuss elaborate treatment plans; at a minimum, plans can be made to meet again at a specific time: “Let’s take a break so that we can consider what this news means and plan on meeting again in an hour [or tomorrow, or next week] to discuss what to do.”
Leave but do not abandon	Be clear about leaving and about the plans to return: “I’m going to go now, and will be back to see you again in an hour.”
Debrief	Discussing bad news is remarkably demanding and difficult work; a quick debriefing with another staff member who was present during the discussion can be useful in considering how effective the discussion was, how your technique might be improved, and what emotional impact the discussion has had on you

another kind of conversation focuses on defining new goals and plans that confront the reality of the bad news. These and other conversations strive to deepen the therapeutic alliance among patient, family, and health care team. A framework for therapeutic planning discussions, based on a simple yet powerful model of decision making, is outlined in [Table 2](#).

Table 2
Framework for therapeutic planning conversations

Steps	Comment
Review the major problems	Develop with the family a shared understanding of the major problems confronting them and their child: "I think we all find it useful to hear what each of us think are the most important problems and challenges that we need to confront."
Discuss goals, objectives, hopes	The compass by which the course of therapy will be set depends on a clear sense of purpose and the values that underlie that purpose; you can initiate this phase of the discussion with the following question: "Given the problems and challenges, I am thinking about how we can best care for your child. What are your major hopes or goals?" Encourage each participant to state all of their hopes or goals, which may not be entirely consistent (such as "to be cured by a miracle" and "to not suffer before dying"); the aim of this phase is simply to air all of the hopes, goals, or objectives and summarize them for the group when everyone has spoken
Spell out alternative actions	In as nonjudgmental and succinct a manner as possible, clarify the main alternative ways of caring for the child, such as: "I feel that we have two major ways we can proceed, namely, continue to have your child stay in the hospital or work to have your child go home with the help of hospice or home-care nursing." Check to see if anyone has another major option
Examine consequences of options	Typically, no therapeutic option is perfect; once the main options are spelled out, explore as a group the pros and cons of each alternative. "If we keep your child in the hospital, there are some good and not-so-good things I can foresee happening, such as . . ." The judgments of "good and not-so-good" should refer back to the discussion regarding hopes and goals; solicit other participants' perspectives
Explore tradeoffs	Often a central pattern of tradeoffs between two goals emerges in the discussion of options; addressing this tradeoff explicitly can be helpful: "We seem to be struggling with the problem that the goal of feeling secure, a sense of which the hospital provides, is at odds with the goal of being at home." Stating these tradeoffs can help people clarify which goal is more important or devise new alternatives to accomplish both goals (eg, enhancing the sense of security at home by augmenting the nursing coverage)
Formulate a plan	By this point, the broad elements of a plan typically have emerged by consensus; if so, spell it out and assess whether everyone agrees; if not, acknowledge the disagreement and seek agreement to continue to work together to formulate a mutually acceptable plan
Specify next steps and follow-up assessment	Before ending the conversation, clarify who will do what next and when the next discussion will occur

The first five steps of this framework can be remembered with the mnemonic 'ProACT' (as in, be proactive): PProblem, Objectives, Alternatives, Consequences, Tradeoffs.

Involving the family

When the goal of care shifts from cure to providing comfort, a family often looks to the health care team for guidance regarding how to provide the best care for their child. Family-centered care is an underlying principle of palliative and end-of-life-care. Families should be encouraged to maintain their lifestyle to the best of their ability and ensure that their belief system is respected. In discussing quality end-of-life care, the health care team must recognize a family's cultural values and traditions.

Conversations with families should emphasize the importance of improving the quality rather than the quantity of a child's days. There are many ways to maximize quality of life for dying children. Because time is of paramount consideration, the focus should be on making each day meaningful. Health care providers should encourage a child and family to make memories of moments big and small and to celebrate "milestones" (eg, first tooth, first words, birthdays, graduations, religious ceremonies) while the child is still able to participate. Activities such as keeping a journal, making a photo album, videotaping special occasions, making imprints of the child's hands or feet, sharing stories, and keeping locks of hair all help create memories for the whole family.

Families also may look to the health care team for advice on how to communicate with their children about death. Although a child may be acutely aware of his or her condition, parents may have difficulty talking with the child about death. It may be helpful for families to know that studies have shown that children are aware of their condition and impending death even when no "official" conversation has taken place. Although we may believe that children have the right to honest answers about their illness, we also recognize that parents are the fundamental decision makers. Often religious, spiritual, and family traditions play an important role in how parents communicate with their children. One way to help parents with these difficult conversations is to ask them what worries them when answering their child's questions about death or illness. One may consider involving the family's spiritual advisor or another member of the health care team (eg, the hospital chaplain, social worker, or child-life specialist) to facilitate such conversations.

Psychosocial and spiritual needs

Dying children and their families have a host of concerns—psychological, financial, spiritual, cultural, and ethical—that good palliative care should address. Psychosocial interventions seek to provide families with a safe environment to express and process the intense emotions during times of crisis, including fear, grief, anger, loneliness, stress, and isolation, and cope with the challenges as successfully as possible [8]. Each member of a palliative care team, including mental health care and medical professionals, can play a role in these interventions. Whereas all psychosocial professionals, including psychologists, social

workers, child-life specialists, and pastoral care workers, tend to focus on issues of adjustment, communication, and making meaning, there is typically a degree of specialization of duty by profession.

Social work with patients at the end of life is multidimensional and can include case management, counseling, advocacy, and acting as a liaison between the patient, family, medical team, and hospice agency [9]. Palliative care social workers can act as a repository for the family's normal but overwhelming feelings of suffering, fear, and loss while facilitating communication between the dying child and his or her family. They also can help families navigate the complex medical and social service systems and access financial support resources while a child is in the hospital and once the child is home. Above all, social workers support and advocate the hopes and wishes of the child and family and remain present with the dying child and family to acknowledge their suffering and help create an environment of hope and healing.

Psychologists can contribute expertise in the areas of formal assessment and intervention, particularly when normal means of coping that usually enable people to adjust to difficult situations become maladaptive. The world of the seriously ill child is often filled with high levels of anxiety and other distress that frequently go untreated in the face of medical needs, which are often seen as more pressing [10]. Psychologists can assess a child's anxiety and distress and then implement formal interventions to reduce troubling symptoms, including serving as a coordinating liaison with psychiatrists. The same kind of assessment can take place on a family level, with psychologists working to assess family needs and provide interventions when distress diverts a family from a more adaptive coping and grieving process. Whereas psychologists are most often called on when psychopathology is noted, psychologists can provide interventions for all families who are confronting serious or terminal illness with the goal of supporting positive or adaptive coping over time [11].

In the hospital setting, child-life specialists and creative arts therapists often function as translators of a child's experience. They search, with the patient and family, for play or art modalities that stimulate imagination and create a bridge or transitional space between a child's inner and outer worlds [12]. Allowing the opportunity for choices in art and play may provide control to patients when they have little choice about being ill or needing medical care at the end of life. Although children and adolescents are not always able to communicate their feelings, art and play can assist them in finding words to describe their experiences and work through traumatic experiences associated with hospitalization, illness, and dying. Creative therapeutic expression can promote a sense of independence and control, facilitate positive self-esteem, and encourage self-expression of thoughts and feelings. By communicating to a patient that all feelings are acceptable, child-life specialists and creative arts therapists develop a therapeutic relationship in which a patient feels accepted and safe. At a time when feelings may be difficult to put into words, art and play can help a child and other family members to express the experience of approaching death and their feelings of impending loss, which provides support to the whole family.

Spiritual support, a critical component of palliative care, is most often the province of pastoral care workers but should be considered by all members of the team. One goal of spiritual care for dying children and their families is to help them find meaning in living and dying. Structured spiritual assessments of a child or family's formal religious and spiritual affiliations, beliefs, practice, and rituals can provide powerful insights, which often lead to intense conversations with the dying patient and family about their search for meaning and what occurs after death. Supportive discussions can help build meaning, offer comfort before and after a child has died, and provide strength during the grieving process. Many dying children and their families wish to have prayers or other spiritually significant rituals occur in the hospital, and pastoral care workers can assist in achieving these goals. Pastoral care workers and social workers also can act as liaisons between the family and outside resources, such as funeral homes, churches, and temples, to ensure that the family's funeral and burial wishes are fulfilled.

Specialization across the psychosocial professions is helpful and necessary. In a mature and effective palliative care team, however, the psychosocial professionals are interdependent, with some overlapping responsibilities and with each professional supporting the roles and responsibilities of the other.

Symptom management

A cornerstone of good palliative and end-of-life care is the alleviation of suffering by the skillful management of symptoms. Several core principles underlie all symptom management strategies. First, patients must be assessed actively and frequently in a comprehensive manner for distressing or bothersome symptoms, including the assessment of "total" pain, which encompasses not only physical pain but also emotional, psychological, social, and spiritual suffering. Second, neither evaluating nor treating the symptom should be more distressing or bothersome than the symptom itself. Finally, if one treatment method is not successful, several different modes of treatment often work synergistically to control a symptom.

Most children experience pain at some point during their illness. Pain is a subjective symptom that is physically and emotionally distressing for a child and the caregiver. Pain can be somatic, visceral, or neuropathic and can be caused by disease-related, treatment-related, or psychological distress. The key to effective pain control is frequent, detailed assessment that includes location, duration, and possible causes. The current standard for pain management in children who have cancer and children who are receiving palliative care consists of four concepts developed by the World Health Organization: (1) by the ladder, (2) by the clock, (3) by the mouth, and (4) by the child. That is, treatment should be escalated according to the World Health Organization Analgesic Step Ladder approach outlined in [Table 3](#), be administered on a scheduled basis, be given by the least invasive route, and be tailored to an individual child's circumstance and needs

Table 3
Escalation of therapy to control pain

Step of therapy	Indication	Guidance
1	Mild pain	Nonopioid analgesics: acetaminophen, ibuprofen, naproxen +/- adjuvants: anticonvulsants for neuropathic pain and antidepressants or anxiolytics for coexisting mood disturbances
2	Moderate pain or mild pain not relieved by step 1	Opioid analgesics for step 2: codeine, oxycodone, morphine +/- nonopioid analgesics +/- adjuvants
3	Severe pain or mild-to-moderate pain not relieved by steps 1 or 2	Opioid analgesics for step 3: hydromorphone, fentanyl, patient-controlled analgesic delivery of intravenous narcotic +/- non-opioid analgesics +/- adjuvants

Based on the World Health Organization "ladder" approach to cancer pain management.

[13]. In addition to pain medications, supportive, behavioral, and cognitive methods for alleviating pain may be appropriate for children.

Many parents fear the use of narcotics and need reassurance from their child's medical team. Physicians also must anticipate and manage side effects of narcotics, such as fatigue, constipation, and nausea. If side effects become intolerable, opioids can be rotated; that is, the patient is switched from one opiate to another opiate, with often a slight reduction in the equianalgesic dosage, so that pain is still controlled but side effects are diminished. Table 4 contains equianalgesic opiate doses.

Table 4
Equianalgesic narcotic dosing

Medication	Parenteral dose (mg)	Oral or rectal dose (mg)
Morphine	1	3
Codeine	12	20
Hydromorphone	0.3	0.8
Hydrocodone	NA	3
Oxycodone	NA	2
Fentanyl patch	1.5 $\mu\text{g}/\text{h}^{\text{a}}$	NA
Methadone	NA	Variable ^b

^a Fentanyl patches are available in 25-, 50-, 75-, and 100- $\mu\text{g}/\text{h}$ doses. Given individual variability in conversion, underestimating the initial fentanyl patch dose is appropriate, augmented with rescue doses of oral morphine if pain is not controlled. The dose of 1.5 $\mu\text{g}/\text{h}$ is equianalgesic to 1 mg of oral morphine taken every 4 hours; a dose of 25 $\mu\text{g}/\text{h}$ would be equianalgesic to 15 mg of oral morphine taken every 4 hours. Patches take 18 to 24 hours to achieve steady-state serum levels of fentanyl and once removed have a half-life of another 18 hours.

^b Methadone offers remarkably effective pain relief but should be prescribed by a physician familiar with this medication and the management of pain.

Nausea and vomiting are a common set of symptoms at the end of life. Causes are multifactorial and can include chemotherapy, central nervous system triggers, and gastrointestinal abnormalities. Because being unable to eat or drink as the result of nausea or vomiting has a significant impact on a patient's quality of life, assessment and treatments should be instituted quickly. Treatment is based on likely cause and can include pharmacologic and nonpharmacologic methods. In cases of chemotherapy-induced nausea and vomiting, medications such as prochlorperazine, ondansetron, scopolamine, and metaclopramide can be effective, as can 5-HT₃ receptor antagonists (eg, granisetron and ondansetron). One small study showed ondansetron to be effective in the palliative care setting for adults admitted with the chief complaint of nausea and vomiting [14]. Multiple agents often are necessary for effective symptom relief. Nonpharmacologic methods include providing smaller, frequent meals, taking medications after meals if possible, and avoiding smells and tastes that exacerbate the symptoms.

Fatigue, one of the most prevalent symptoms in children at the end of life, is generally defined as the subjective feeling of being tired and lacking energy, which can be caused by depression, anxiety, pain, poor nutrition, medication side effects, hypoxia, infection, and dehydration. Although there are several assessment tools for fatigue in children, diagnosis is based on subjective report by the patient or parent [15,16]. As with nausea and vomiting, treatment seeks first to remedy any possible underlying causes. Maximizing a patient's rest also is important and may involve the use of sleep agents. Pharmacologic and nonpharmacologic interventions have been used in managing cancer-related fatigue in adults. Psychostimulants have been used in patients who have cancer and patients who do not have cancer patients for the treatment of fatigue, especially in patients who have been sedated with opioids. Amphetamines have been used in multiple trials in adult patients who have cancer, HIV, chronic fatigue syndrome, and multiple sclerosis. In adults, psychostimulants, such as methylphenidate or dextroamphetamine or corticosteroids, may alleviate fatigue [17–20]. Although psychostimulants are generally well tolerated and may improve mood, energy, and cognition, in children they can cause jitteriness, insomnia, and decreased appetite. In general it is best to give the medication in the morning and early afternoon rather than in the late afternoon or evening. Nonpharmacologic methods include eliminating sedating medications; establishing a sleep schedule with rest periods throughout the day; maximizing nutrition and hydration by means that accord with patient wishes; and engaging in mild exercise as tolerated.

Anxiety, which is commonly experienced by children who have life-shortening conditions, can be a normal response to several factors: painful procedures, uncomfortable symptoms, undesirable medication side effects, the progression of disease or worsening impairment, diminished choices or loss of control, separation from friends or family environment, and other predicaments that arise in the course of their illnesses. Sometimes, the specific causes of a child's anxiety can be identified and treated effectively. When no specific treatable cause can be uncovered, the most common therapeutic approach involves either an oral or an intravenous benzodiazepine, such as lorazepam, diazepam, or

midazolam. This class of medication is known to cause sedation, however, and there is little evidence that the medications effectively diminish anxiety *per se*.

Constipation, although subsequently recalled by parents of dying children as one of the major bothersome symptoms, is often underrecognized and treated ineffectively by physicians. Reduced oral intake of food or liquid, diminished physical activity, and the use of constipating drugs such as morphine or the other opiates, along with potential gastrointestinal dysmotility as a result of the disease lead to constipation. Prevention of constipation is ideal, with daily attention to whether a patient has had a recent bowel movement combined with daily administration of a stool softener (eg, polyethylene glycol or docusate) and a bowel stimulant (eg, senna). Despite best efforts, constipation sometimes may require treatment with an enema, but such events should prompt a thorough re-evaluation of the preventive medication regimen and possible escalation of dosages.

Dyspnea is the sensation of breathlessness that occurs when the body is unable to meet its ventilatory need. A difficult complaint to quantify accurately, dyspnea is important to recognize and treat, because it can be uncomfortable and herald the onset of respiratory failure [21]. Treatment first should target any and all underlying causes and seek to rebalance ventilatory capacity and metabolic demand. Obstructive airway disease or restrictive lung disease can burden the respiratory system, and respiratory muscle fatigue or limitation because of pain can diminish ventilation to the point at which the respiratory system no longer satisfies the body's basic metabolic needs. Alternatively, the body's metabolic demand may increase beyond what the ventilatory capacity of the lungs can support. For patients with significant tachypnea and obstructive lung disease, bronchodilation and anti-inflammatory therapy should be the first line of treatment. If the dyspnea continues, the physician is obliged to discuss goals of care with the patient and family, explaining possible levels of intervention, such as noninvasive continuous positive airway pressure, bilevel positive airway pressure, or mechanical ventilation. Supplying continuous positive airway pressure may help to prevent premature airway closure during exhalation and increase inspiratory flow, thus alleviating a patient's sensation of not being able to inhale deeply enough. Inspiratory pressure augmentation using bilevel positive airway pressure can help to expand the chest wall, decrease the work of breathing, and improve ventilation in patients with restrictive lung disease, thus reducing dyspnea. Oral and parenteral opiates (eg, morphine) and anxiolytic agents (eg, benzodiazepines) can be titrated upward until the sensation of dyspnea is relieved. Although inhaled narcotics and analgesics have been used to palliate dyspnea in an attempt to minimize the respiratory depression and sedation that systemic opiate treatment can cause [22], the results are mixed [23]. Nebulized morphine has been used in most of the published studies, although nebulized fentanyl has been used successfully [24]. Finally, calming techniques, hypnosis, and the use of fans blowing air across a patient's bed may help increase comfort and minimize dyspnea [25].

Seizures, which are common at the end of life, occur as a result of progression of the primary central nervous system disease or of progression of the

primary disease to the central nervous system. Seizures also can result from high fever or metabolic derangement (most frequently hypoglycemia, dysregulated sodium homeostasis, and hypocalcemia). Managing seizures at the end of life is critical, because they can create a barrier between a patient and the family. Parents who have witnessed a child convulsing commonly state that they never want to see their child go through a seizure again [26–28]. Although patients seem to be in agony during a seizure, they report few physical symptoms—confusion, muscle aches and pain from local traumas—after an episode. They are more likely to fear future mental handicap [29] and embarrassment about losing control of consciousness and especially bladder or bowel function in front of friends and family during a seizure [30].

In general, identifying the cause of the seizure is of paramount importance, because some metabolic causes cannot be treated until the underlying cause is corrected. In cases in which a patient is being cared for at home or on whom blood draws have been limited, seizures may be the life-ending event. Various factors influence clinical decisions regarding medications for treating seizures: the acuity of presentation, the presence of other organ damage or malfunction, and, in the context of end-of-life care, the wish to preserve or cloud patient consciousness.

For the acute treatment of a seizure, intravenous benzodiazepines are the therapeutic mainstay. Rectal valium is an excellent alternative when the therapeutic aim is to stop a seizure. For the ongoing management or suppression of a seizure disorder, benzodiazepines can be given on a scheduled basis, but this practice should be avoided unless death is imminent because benzodiazepines cause substantial sedation and may depress respiration. Valproic acid causes negligible sedation but should not be used in patients with mitochondrial defects or other inborn errors of energy metabolism, in children who are younger than 2 years who may have unrecognized metabolic disease, patients who have bleeding diatheses (because it interferes with platelet function), or patients who have liver dysfunction. The best long-term nonsedating options may be the newer anticonvulsant agents, such as levetiracetam or oxcarbazepine. Both medications are available only in oral formulations (liquid and tablet) and achieve good serum concentrations after only two doses, with good overall tolerance. The primary side effect of levetiracetam is behavioral change [31]. Oxcarbazepine, which may cause some initial sleepiness, is also effective for neuropathic pain [32].

Hospice and home nursing care

Not all families choose to care for their dying child at home. For families who do, however, having their child at home during their final days or months can give them a sense of fulfillment and comfort. When discussing the option of pediatric hospice with families, it is helpful to dismantle some misconceptions and answer any questions the family may have regarding hospice service.

The decision to go home on hospice care requires advanced planning and coordination with the health care team, the hospice provider, and the family. To make the transition as seamless as possible, the health care team must have a well-organized plan that is communicated to everyone involved. As an advocate for the child and family, the health care team can help identify appropriate services and providers and clarify the treatment plan and patient needs with the third-party payer to maximize the child's health care benefits.

Although hospices were developed using an adult model of care, many hospices have the capacity, skill, and desire to care for children. The appropriateness of a particular hospice agency can be determined by asking certain questions:

- Has the agency had experience in caring for pediatric patients?
- If so, what was the experience like?
- Do they have staff who have been trained in end-of-life care for children?
- Are they available to come to the hospital to meet the family before discharge?
- Can they provide ongoing bereavement services after the child dies?

Once a hospice program has been identified, it is valuable to discuss with the family their fears and concerns about hospice. Because many families equate hospice with “going home to die” or “giving up hope,” they need to know that choosing hospice does not mean forgoing options, such as returning to the hospital or continuing care with their primary physician. Most pediatric hospices do not require parents to sign a do-not-resuscitate order. Any conversations the primary care physician has had with the family about limits of care should be shared with the hospice provider, however.

The main goal of pediatric hospice is to enhance a child's quality of life in his or her final journey. We must remember that the journey belongs to the child and family and that the job of the multidisciplinary health care team—and the partnership between hospital and hospice—is to support them on that journey.

Bereavement

Although the generic process and parameters of grief are well established in the literature [33], the actual experience of loss and adjustment takes different forms. The process of grief is not linear and never fits neatly into predetermined categories and time scales. The way in which support services are provided to families affected by the death of a child and the way in which they are involved before and after the death is crucial to how they experience the loss, are able to accept it and, subsequently, work through the bereavement process. When medical care can extend life for increasingly longer periods between diagnosis of a terminal illness and the actual death, families often have time to anticipate the loss of their child and experience anticipatory grief. This is an important

part of the grieving process, one in which family members should be encouraged to acknowledge their feelings and speak openly about the imminent death.

Soon after a child dies, the family members left behind experience acute grief. A health care professional's presence during this time is an important way to offer support and comfort. Most families express the feeling that it helps when people acknowledge the significance of their loss. The health care providers who treated the child should share memories with the family and thank them for the opportunity to have participated in the child's care. They should focus the discussion on the sadness of the death rather than the clinical details of the illness and death. They can provide the family with printed information on grief or refer to community organizations or agencies for support and counseling. By giving family members the information they need, an opportunity to talk about their feelings, and suggestions for coping, the pediatrician can help everyone concerned cope somewhat better with the tragedy of a child's death.

Many physicians and other health care professions experience caregiver grief, which is expressed as feelings of failure, sadness, or frustration, when a child they are caring for dies. Honesty and genuine expression of emotion to others allows a health care provider to be more sensitive to those in your care. By being able to recognize your own grief and responses to grief, you will be able to seek support yourself and be more effective in offering support to others.

A comprehensive bereavement program helps families start on the painful journey of grief. Bereavement services should be tailored to meet the unique needs of families. Grieving families should be provided with alternatives and encouraged to choose which services, if any, suit their needs. Bereavement follow-up services may include individual counseling, support groups, memorial events, written resources, and education. Grief counseling can be helpful for people who are grieving or anticipating the death of a loved one. Professional counseling provides a safe place for expressing and normalizing feelings of grief. Although the journey is arduous, offering support and guidance can help families begin the necessary process of healing.

When parents who have had a child die meet together in a grief support group, a sense of peace and healing can result. Many parents find comfort knowing that they are not alone in their grief. Various types of community-based grief support groups are available. Many hospices, hospitals, religious institutions, and funeral homes offer bereavement support groups that are open to all members of the community.

Ethically problematic situations

Pediatric hospitalists must be able to recognize and manage ethically problematic situations, often by drawing on other personnel and resources at their hospitals. This section presents three common situations encountered in the realm of palliative and end-of-life care [34].

Scenario 1: truth telling

You are caring for a 12-year-old girl with rapidly progressive glioblastoma multiforme who presented with a right hemiparesis. Unfortunately, she is not a candidate for any curative treatment and has a poor prognosis. Every time you see her, she asks what is wrong with her and when she will be able to walk again. Her parents prefer to let her think she has a virus rather than telling her the reality of her diagnosis and prognosis.

In pediatrics, most decisions are guided by the best interest principle [35]. This situation presents a challenging conflict between truth telling and denial because there are reasons to justify both courses of action [36]. Arguably, the child's parents think her interests are best served in not knowing that she has a terminal, rapidly progressive illness. From the child's viewpoint, however, she might want to be informed so that she can express her feelings openly and have an honest conversation about dying. This kind of situation is particularly challenging because health care providers often feel a duty to the patient to be honest. They should not, however, disclose her illness and prognosis to her without the family's knowledge. Although there is a paucity of data about the benefits of truth telling with terminally ill children, the American Academy of Pediatrics reports that children and adolescents who know that they have HIV have greater self-esteem than do those who do not know their status [37].

In this case, it is best to counsel the family on the importance of telling their daughter the truth, especially in light of her repeated questions. Child-life services and psychosocial support should be offered to the patient and family. If a conflict between the health care team and the parents persists, any member of the health care team may bring the case before the institutional ethics committee for help in resolving the issue.

Scenario 2: futility

You are the pediatrician on call in a level II neonatal intensive care unit when a full-term baby with a prenatal diagnosis of trisomy 18 is born. Over the ensuing days, the baby has multiple episodes of apnea that require resuscitation with bag mask ventilation. You talk to the family about possible do-not-attempt-resuscitation status and present the option of providing the baby with feeds and palliative care but not cardiopulmonary resuscitation. The parents state that they want everything done because the baby's grandparents told them that was the law.

The grandparents likely recall the outcry over the federal Baby Doe regulations, which were promulgated in 1982 after a baby with Down syndrome and a trachoesophageal fistula died without surgical intervention [38]. The regulations required that all newborns, even those with terminal illnesses, receive maximal life-prolonging treatment. A hotline phone number was established for hospital staff to report cases anonymously in which care did not conform to the rules. In reality, however, these regulations (currently codified in the federal Child

Abuse and Neglect Act) never have been enforced, nor do they have the power of law or regulation to affect individual level decision making.

The fundamental conflict lies in the parents' insistence that the baby receive cardiopulmonary resuscitation and the health care team's perception that this intervention cannot succeed and represents futile treatment [39]. Currently, however, there are no universally accepted guidelines regarding what constitutes futile treatment, and unilateral do-not-attempt-resuscitation orders are not acceptable. Instead of arguing about whether a treatment is futile, alternative avenues to resolve disputes about limiting resuscitation should be pursued, including perhaps an ethics committee consultation or mediation regarding a do-not-attempt-resuscitation decision [40].

In this case, the primary physician should arrange a family meeting to clarify the parents' goals and hopes for their baby. Perhaps the parents want the baby resuscitated so he can live just long enough to partake in a religious ceremony or meet an important family member. Perhaps they want the baby to live for months or years and will pursue a tracheotomy and mechanical ventilation to care for the baby at home. Clarification of these goals or hopes can help health care professionals understand and empathize with patients or families and move from conflict to collaboration.

Scenario 3: withdrawal of artificial fluids and nutrition

A 10-year-old girl with progressive neurologic failure from an underlying metabolic disorder is admitted with increasingly frequent seizures. Over the last 6 months, she has lost her ability to take anything orally. During her hospitalization, the parents tell you that they feel they are only prolonging their child's suffering. They have talked to their pastor and have decided they would like to stop gastrostomy tube feeds and want their daughter to receive seizure medication and comfort measures only.

This is a challenging situation, because the withdrawal of fluids and nutrition delivered by "artificial means" is emotionally, morally, and politically charged. Ethically, fluids and nutrition provided by intravenous administration or delivered by a feeding tube are considered a form of life-sustaining medical treatment. With parental agreement, physicians may forgo giving hydration and nutrition in cases in which they believe such measures are not "appropriate" [41]. In this case, the physician should try to involve health care providers who have had a long-term relationship with the family to explore whether the parents' decision is based on their concerns about prolonging suffering and inadequate seizure management or whether it is a result of their own exhaustion and need for respite. The family and the health care team must believe that this decision is in the child's best interests. If, after consulting with the primary pediatrician and neurologist, the consensus is that continuation of artificial fluids and nutrition is only prolonging the patient's suffering, then withdrawing artificial fluids and nutrition and instituting aggressive comfort-care measures would be appropriate.

Improving end-of-life care

Regardless of one’s degree of experience in pediatric palliative care, improvement is always possible and imperative. Individual health care providers and teams of providers can gain a clearer sense of how to improve the care that they provide by periodically undertaking a self-assessment that considers the major quality-of-palliative-care domains. Table 5 presents a framework for this self-assessment, along with sample questions.

After identifying domains in which significant improvement is possible, individuals or teams should target one area at a time for study (various excellent resources are available in print and on the worldwide web) or discussion and planning with other members of the hospital staff (eg, physician colleagues, nurses, chaplains, psychologists, or members of the hospital administration). Step by step, we can improve the care received by dying children and their families.

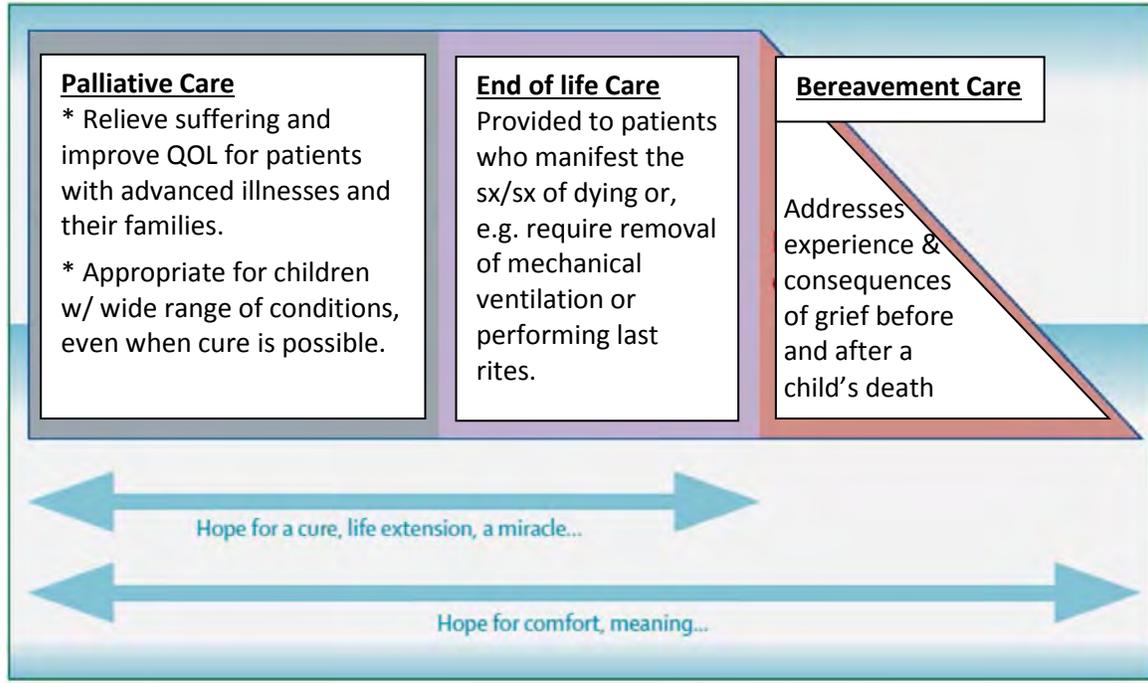
Table 5
 Framework for assessing and improving quality of care

Quality-of-care domain	Self-assessment questions
Collaborate, communicate, and support decision making	How competent do I feel in these areas? What feedback can other staff members give me about how I am performing?
Minimize bothersome symptoms	Am I doing a good job of pain assessment and control? Is there a particular symptom that I feel uncomfortable managing?
Provide emotional and spiritual support	Whom should I ask to help me provide these kinds of support? Social worker? Child-life specialist? Psychologist? Pastoral care staff?
Maximize other quality-of-life enhancers	Do I have a sufficiently clear sense of what this child and family value most to make sure that our plans support these values? Have I forged ties with all possible allies, from the child’s friends to school or wish-granting organizations?
Institute palliative care in a timely manner	Can I raise the issue of symptom management and quality-of-life enhancement earlier in the course of illness to prepare for subsequent conversations?
Visualize and address the full population at need	Are all the children I care for who might die as likely to receive quality end-of-life care? If not, why not?
Provide a continuum of care across multiple sites	How can I improve communication and transitions of care between in-patient and out-patient settings?
Appreciate and manage tradeoffs adroitly	Am I sensitive to the various difficult tradeoffs that patients or parents struggle with or that I am struggling with? How well am I recognizing and managing ethical dilemmas when they arise?
Operate in accord with an evidence base to maximize safety and effectiveness	What do I know about the drugs or other interventions that I am using? Can I find published systematic reviews to enhance my understanding of effective treatment?
Practice the art of individualization	How can I forge more effective therapeutic alliances with specific children and their families?
Provide self-care	Am I acknowledging the impact that caring for dying children has on me? What am I doing to care for myself in this role?



Palliative Care Quiz

1. Complete the following diagram of the **Stages of Palliative Care** and define each stage: (from *Lancet* 2008; 371: 852-64) :



2. List **4 pediatric conditions** for which palliative care is appropriate? *Do you have any patients who are receiving (or should be receiving) palliative care?*

Table 1. Conditions Appropriate for Pediatric Palliative Care.*

<p>Conditions for which curative treatment is possible but may fail Advanced or progressive cancer or cancer with a poor prognosis Complex and severe congenital or acquired heart disease</p> <p>Conditions requiring intensive long-term treatment aimed at maintaining the quality of life Human immunodeficiency virus infection Cystic fibrosis Severe gastrointestinal disorders or malformations such as gastroschisis Severe epidermolysis bullosa Severe immunodeficiencies Renal failure in cases in which dialysis, transplantation, or both are not available or indicated Chronic or severe respiratory failure Muscular dystrophy</p> <p>Progressive conditions in which treatment is exclusively palliative after diagnosis Progressive metabolic disorders Certain chromosomal abnormalities such as trisomy 13 or trisomy 18 Severe forms of osteogenesis imperfecta</p> <p>Conditions involving severe, nonprogressive disability, causing extreme vulnerability to health complications Severe cerebral palsy with recurrent infection or difficult-to-control symptoms Extreme prematurity Severe neurologic sequelae of infectious disease Hypoxic or anoxic brain injury Holoprosencephaly or other severe brain malformations</p>
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Table from NEJM 2004; 350: 1752-1762

3. What are the different **types of grief**?

- (a) Anticipatory grief: Occurs during the time between diagnosis of a terminal illness and the actual death, as families. Family members should be encouraged to acknowledge their feelings and speak openly about the imminent loss.
- (b) Acute grief: Occurs soon after a child dies. Providers should share memories with the family and focus on the sadness of the death, rather than the details of the illness.
- (c) Caregiver grief: Feelings of failure, sadness, or frustration when a child they are caring for dies. Recognizing one's own grief can help providers offer support to others.

4. Complete the following table for **symptom management** in palliative & end-of-life care.

Remember that many of the treatments may depend on a family's wishes for extent of care:

Symptom	Causes	Pharmacologic Rx	Non-pharmacologic Rx
Pain	Disease-related Treatment-related Psychological	Non-opioid analgesics → opioids ± adjuvants	Supportive, behavioral, and cognitive methods. (Child Life, Psych, S/W)
Nausea & vomiting	Chemotherapy CNS triggers GI abnormalities	Zofran, Kytril, Reglan	Small, frequent meals Meds after meals Avoiding triggers
Fatigue	Psych, pain, poor nutrition, meds, hypoxia, infection, dehydration	Sleep agents Psychostims (Ritalin) Steroids	Maximize rest- sleep schedule Eliminate sedating meds Mild exercise, good nutrition
Anxiety	Procedures, sx's, med side effects, disease progress, lost control, family separation	PO or IV benzodiazepine	Supportive, behavioral, and cognitive methods.
Constipation	↓PO; ↓activity; opioid side effects; GI dysmotility	Stool softener (Miralax, Colace) + stimulant (senna); enema	Rotate opioids, with slight reduction in equianalgesic dose; NG feeds.
Dyspnea	-Obstructive or restrictive lung dz. -Muscle fatigue or limitation from pain. -↑ Metabolic demand.	-Bronchodilators; corticosteroids. -CPAP, BiPAP, MV. -Opiates and anxiolytics relieve sensation.	Calming techniques, hypnosis, fan blowing air across bed.
Seizures	1 ^o or 2 ^o CNS disease; fever; metabolic derangement	Treat etiology IV or rectal benzos; Keppra, Trileptal	None.

Bonus: What is the WHO standard for pain management in children receiving palliative care?

(1) By the ladder (see **Tables 3 & 4**); (2) by the clock (*scheduled*); (3) by the mouth (*least invasive route*); and (4) by the child (*tailored to child's condition and needs*)